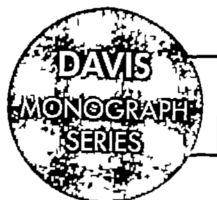
CARDIAC SURGERY
1960-61

Edited by CHARLES P BAILEY, M D

Chairman and Professor of Surgery New York
Medical College, Flower and Fifth Avenue Hospital
New York N Y



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Foreword

This monograph comprises a series of articles dealing with the surgical therapy of the more common cardiac lesions Stress is placed on the aspects which have been developed more recently This is particularly true of the chapters which deal with the surgery of acquired heart disease

No attempt is made to cover the entire subject, but rather to highlight the considerable progress which has been made in this field In addition to the basic philosophy and concepts it also includes detailed description of present-day operative technics, which would seem to be of value for the foreseeable future

Considering the speed with which the field of cardiac surgery is developing, as well as the fact that it is being practiced more and more widely all over the world, it would seem most timely to present this collection of articles which represent the experience and practice at two of the larger Clinics

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CHARLES P BAILEY, M D

Contributors

HOUCK E BOLTON M D

Surgeon Edgewater Hospital Chicago Ill

DANIEL F DOWNING M D

Associate Professor of Pediatrics, Hahnemann Medical College and Hospital, Philadelphia, Pa.

EDWARD A FITCH M D

Surgeon, Cardiovascular Section Department of Surgery Hermann Hospital Houston, Texas.

WILLIAM M LEMMON M D

Associate in Surgery New York Medical College Flower and Fifth Avenue Hospital Assistant Visiting Surgeon Metropolitan Hospital and Bird S Coler Memorial Hospital and Home New York Thoracic Surgeon Deborah Hospital, Browns Mills New Jersey

DRYDEN P MORSE M D

Director of Thoracic and Cardiovascular Surgery Passaic General Hospital, Passaic, N J Director Cardiac Surgical Unit St. Francis Hospital Trenton, N J Instructor and Assistant Surgeon, Department of Thoracic Surgery Hahnemann Medical College and Hospital Surgeon Deborah Sanatorium, Browns Mills N J Assistant Chief Thoracic Surgery Philadelphia General Hospital Surgeon Woman's Hospital, and Albert Einstein Medical Center Philadelphia, Pa.

JACOB ZIMMERMAN F R C S

Research Fellow in Surgery New York Medical College, New York, N Y

Contents

COARCTATION OF AORTA

WILLIAM M LEMMON M.D., AND CHARLES P BAILEY M D

Coarctation of the aorta amounts to a narrowing or stricture of this vessel at some point in its course In 98 per cent[1] of the cases the lesion is located just distal to the origin of the left subclavian artery The severity of the obstruction usually is sufficient to produce alterations in the normal physiology which are detectable on routine physical examination. They are manifested by a basal systolic heart murmur elevation of the blood pressure in the upper extremities, and an absence of the normal differential in the arterial pressure levels in the arms and the legs.

Classification

While the division of these lesions by Bonnet in 1903[2] into "infantile" and adult" forms probably was acceptable at that time, his method of classification obviously is inadequate in the light of present knowledge of the lesion.[3] The infantile type has been encountered in adults and the adult form in infants.[4]

To satisfy the obvious need for a suitable classification of the lesions we present a simple but complete system of classification[5, 6] (*See also* Fig 1 A E.)

TYPE I

Classic coarctation with ligament (obliterated ductus arteriosus)

TYPE II

Classically located coarctation with shunt.

A. Ductus arteriosus patent to upper aortic segment with left-to-right shunt.

B Ductus arteriosus patent to distal aortic segment with right-to-left shunt.

These shunts should be expressed in liters per minute and may be bidirectional or may become reversed (and should be so designated)

TYPE III

Coarctation of the aorta with hypoplasia (usually of the entire descending aorta)

TYPE IV

Isthmus interruption. Pathologically this type differs from true coarctation in that a segment of the aorta is absent. Physiologically it is similar to Type II B coarctation, a patent ductus arteriosus transmitting pulmonary artery venous blood to the lower aortic segment.

TYPE V

Coarctation occurring in other than usual location.

A. Ascending aorta (no recorded living case)

B Aortic arch.

C. Descending aorta (including the abdominal portion)

D Multiple sites of occurrence.

Etiology

Nearly all cases of coarctation of the aorta are considered to be of congenital origin but the cause of the lesion remains unknown. Embryologists believe that in some instances there is failure of accurate fusion of the dorsal aorta at the isthmus, just distal to the origin of the left subclavian artery and the point at which the ductus arteriosus joins the aorta. Less commonly there is failure of fusion of two embryonic dorsal aortic segments distal to the origin of the renal arteries.[7] However there is no single adequate explanation for these malformations, especially since they can occur at almost any point along the course of the vessel A summary of the various theories concerning the origin of coarctation of the aorta may be found elsewhere.[8]

further decrease the longevity[13] Pregnancy complicated by coarctation of the aorta carries considerably greater risks than usual.[14]

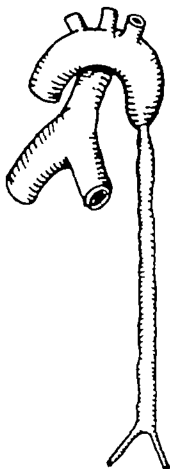


Fig. 1: Type III

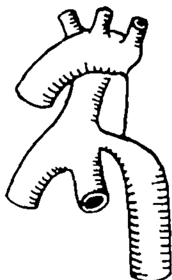
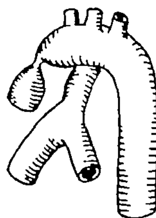


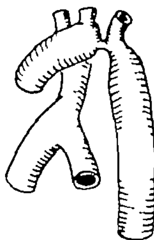
Fig. 1: Type IV

Pathologic Physiology

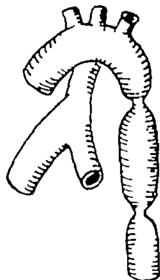
In Type I lesions hypertension usually is found in that portion of the body supplied by the proximal segment. Conversely hypotension commonly exists in the areas supplied by the distal segment. However a series of cases has been observed and reported by Sandifer[15] in which there was normotension in the upper extremities and hypotension in the lower extremities. During fetal life a vast collateral circulatory network is built up about the coarcted



A



B



C

Fig. 1 Type V

segment involving chiefly the tributaries of the two subclavian arteries The anastomosis of the internal mammary arteries via the fourth through seventh intercostals with the distal segment of the aorta and the collateral pathways by way of the lateral thoracic, the transverse scapular, and the anterior spinal arteries are of primary importance. Obviously since the site and degree of obstruction of the lesion is variable, the course and extent of the collateral circulation will assume an appropriate pattern in each given case

Type IIA coarctations usually present a similar collateral system and there is auscultatory evidence of a concomitant patent ductus arteriosus with the usual left-to-right shunt[16] The "machinery type" murmur heard best in the anterior second left interspace readily may be differentiated from the basal systolic murmur of the coarctation The characteristic hemodynamic changes may be elicited by cardiac catheterization

In contrast to the two foregoing types, Type II B and Type IV lesions usually show no evidence of development of a collateral circulation, the portion of the body supplied by the lower aortic segment being nourished by the undersaturated pulmonary arterial blood which flows through the communicating patent ductus arteriosus (right-to-left shunt) Such patients may not have upper extremity hypertension and the lower extremities will be found to maintain blood pressures approaching pulmonary arterial levels If the observer will compare the color of the fingernails with the toe-nails in these patients the cyanosis which is limited to the portions of the body below the brim of the pelvis can be demonstrated readily

Patients with Type III coarctations usually present the same clinical manifestations as Type I patients. Apparently the distal hypoplastic aorta provides sufficient blood flow for survival although one would expect the incidence and the importance of the renal element in the hypertension to be increased considerably.

Type V lesions are a nonhomogenous group with variable evidences of their anomaly, the findings being dependent upon which variant is present

Diagnosis

The presence of aortic coarctation can be ascertained only if the examining physician suspects it and then performs a careful physical examination. Hypertension demands an explanation If this finding is associated with a systolic murmur the examiner is obligated to palpate the arteries of the lower extremity and to compare the color of the hand with the foot, and the radial with the femoral arterial pulsations. In coarctation there is a detectable lag in the pulse wave between these two vessels Upon determination of the blood pressure in the lower extremity an absence or reversal of the normal pressure differential may be found (normally the systolic pressure in the legs is about 40 mm. higher than that in the arms). Such a finding is pathognomonic of coarctation of the aorta

Laboratory Studies: An important part of the preoperative evaluation of every patient in whom a tentative diagnosis of coarctation of the aorta has been made is the electrocardiogram. This tracing will not establish the diagnosis but is essential for evaluation of the status of the myocardium and the conduction system prior to operation Infants and children may not show any abnormality but somewhat later a left axis deviation develops In the older age groups (adolescence or adulthood) usually there is evidence of left ventricular

hypertrophy and occasionally of strain or a frank damage pattern. Bundle branch block is encountered frequently

The radiologic examination in the infant or child *may contribute little or* nothing to the diagnosis. However, there may be cardiac enlargement and, if decompensation has set in, evidence of congestive failure. The cardiac silhouette may be "boot-shaped" suggesting predominant left ventricular enlargement while the aortic arch proximal to the obstruction may be dilated and enlarged. Absence or diminution in the size of the aortic knob may be strikingly evident. In the majority of adult patients there is "notching or "scalloping" of the undersides of the ribs below rib three. This seldom involves all twelve ribs. While not often seen in infants or small children it has been reported Rib notching was formerly considered to be pathognomonic of coarctation of the aorta but, as pointed out by Neuhauser[17] this may be found in patients with intercostal neuromata (neurofibromatosis) longstanding superior vena caval obstruction, the tetralogy of Fallot, or multiple aneurysms of the intercostal arteries.

Esophageal examination by opacification very frequently will reveal, in the posteroanterior view an irregular filling defect on the left side resembling an "E" or reversed numeral 3 due to dilation of the intercostal arteries and impingement upon the esophagus. In the lateral view these same indentations may be demonstrated on the posterior esophageal wall.

While angiocardiography may serve to delineate the beginning of the stricture segment it usually will not indicate the entire extent of it. Since angiocardiography *is not as reliable as supra* sternal transthoracic aortography[18] or retrograde aortography many authorities do not consider it necessary as a routine procedure and when contrast visualization is indicated prefer one of the latter procedures. More important, from the surgical standpoint, is the *status of the aorta immediately* distal to the coarctation for it is here that unsuspected pathology (aneurysm, hypoplasia) or other unrecognized findings may exist. The presence of an unusually long area of coarctation or aneurysm formation of the distal aorta may necessitate *the use of an arterial homograft* or a prosthetic tube for the reestablishment of aortic continuity

Patients who present blood pressure findings other than the characteristic ones, aberrant esophageal signs on barium swallow or other peculiarities in their work up may be suffering from one of the rarer types of coarctation of the aorta and therefore deserve aortographic examination.

Indications for Operation

It is now generally conceded that the mere anatomic existence of a coarctation of the aorta of physiologic significance (blood pressure less in the lower extremities than in the arms) is sufficient justification for surgical correction. The chief controversy devolves upon the optional age for intervention. Probably five to ten years of age embraces the ideal span. The surgical problems with Type I or Type IIA lesions as a rule are clearcut but Types II B III IV and V require careful individual evaluation and treatment.

Contraindications to Operation

1 The presence of active rheumatic carditis, or acute or chronic endocarditis require delay of operation. Medical therapy should *have controlled the* rheumatic process or the infection for at least three months before operation is contemplated.

2 The presence of congestive heart failure in infancy provides theoretical justification for immediate operation to relieve the block.[19] However, the operative mortality under such circumstances is high and many authorities maintain that the best results are obtained by persistence with medical treatment until a more opportune time arrives for intervention [20] In the adult age groups the development of congestive heart failure is indicative of a severely damaged myocardium which may preclude a successful surgical outcome.

3 Advanced age produces a thinned out, inelastic, and often fragile arterial wall with which to work Aneurysm formation at the cephalic end of the distal aortic segment or the proximal portion of the intercostal arteries is not uncommon in such patients Youngsters tolerate operation much better than adults and provide the surgeon with much more satisfactory material for operative repair Age is certainly not an absolute contraindication to surgery, but it may be in the individual patient

4 If an asymptomatic infant or child is found to have a coarctation of the aorta it is accepted practice to observe the patient until he is five to ten years of age at which time operative repair may be undertaken under more nearly ideal conditions, provided complications have not set in If difficulties should arise during the period of observation, operation may be performed at once The children in this "ideal" age group are better able to cooperate, to follow instructions, and are somewhat better operative risks They also have larger and stronger tissues than infants Growth at the line of anastomosis has been proven experimentally and clinically to take place at a normal pace if an interrupted suture technic is used [21]

5 The presence of certain congenital anomalies such as hydrocephalus and mongolism, which have a rather hopeless outlook, suggest the impossibility of worthwhile salvage and may represent an acceptable contraindication to surgery in a given case

Surgical Treatment

The accepted operation of excision of the coarcted segment followed by end-to-end anastomosis of the cut aortic ends is applicable in the vast majority of patients afflicted with this anomaly In 98 per cent of patients with coarctation of the aorta the involved segment lies just distal to the left subclavian artery and is short enough to be corrected by clamping the aorta above and below, removing the stricture, and re-establishing arterial continuity by direct suturing Type I, IIA and Type III cases usually may be handled in this manner

Type II B and Type IV lesions differ in that they lack the collateral circulation necessary for long periods of surgical interruption of aortic blood flow If the arterial flow to the lower part of the body is interrupted for more than fifteen to twenty minutes in such cases the spinal cord is very likely to suffer irreparable damage. Both the mortality and morbidity are high with such management Therefore it is essential to operate on these patients either with the aid of a by-pass shunt pumping left atrial blood to the lower portion of the body, or under hypothermia[22] the reduction in tissue metabolism then permitting a longer permissible period of circulatory interruption

While Type IV patients are similar physiologically to Type II B cases, many of them will require some type of replacement graft or prosthetic tube to bridge the long arterial defect This may be accomplished best by use of the shunt pump The ever-present patent

ductus arteriosus must be closed simultaneously in each of these two types of coarctation.

The surgical therapy of patients with Type V lesions must be carefully planned and individualized. Patients suspected of having Type V D coarctation (multiple sites of stenosis) probably should have both antegrade and retrograde aortographic studies to localize the lesions accurately although contrast studies do not invariably reveal their full extent.

If the coarctation is so long that resection and direct end-to-end anastomosis is impossible or too risky to attempt, several alternatives are available. In those institutions in which circulatory by-pass is readily available it would seem wise to have the apparatus ready as a standby at the time of operation, in addition, a supply of synthetic or homologous arterial grafts should be at hand for immediate use if needed.

The less complicated approaches to the problem should be mentioned

1 The surgeon may retreat, the operative attack being abandoned, and the thorax closed. We have seen two such patients originally operated elsewhere. In both we later performed completely corrective surgery with excellent results.[6]

2 If the operator is unable to reestablish aortic flow by direct anastomosis once the lesion has been excised, the ends of the aorta may be closed. While such an outcome might be life saving no physiologic benefit would be forthcoming It is of interest historically to note that the first definitive operative attack upon a clinically recognized coarctation (with associated aneurysm) was such a procedure.[23]

3 Blalock and Park [4] first proposed turning down the distally detached left subclavian artery to bridge the gap between the proximal and distal segments but Clagett[25] was the first to apply this procedure clinically

4 Later Johnson and Kirby[26] described several methods of enlarging the narrowed upper segment to achieve a satisfactory anastomosis. The subclavian artery was used in most instances with or without an arterial homograft.

5 While excision of the maximally strictured segment may be followed by restoration of continuity by anastomosis conveniently within the fusiform terminations, the resulting reduced calibre of the lumen in these areas may leave something to be desired. However, the poor cosmetic appearance does not always indicate the physiologic result. Wiggers[27] has shown that aortic flow is adequate until the lumen is encroached upon by more than 55 or 60 per cent.

6 The aortic lumen may be enlarged by still another procedure based upon the Z plasty principle as described by Holman.[28]

The first successful employment of a preserved homologous aortic graft to bridge the defect after removal of a long coarcted area was reported by Gross.[29] He also recommended the use of preserved and stored homologous grafts when the aorta is so inelastic that the surgeon is unable to reapproximate the cut ends without undue tension on the suture line. Type IV cases (interruption of the isthmus) can be expected routinely to require use of a replacement "graft of one kind or another In adult patients with coarctation (the lesion obviously has been of long standing duration) it is not uncommon to find poststenotic dilatation of the proximal portion of the distal segment. This may amount to actual aneurysm formation. Such a lesion may necessitate such wide excision of the stricture and aneurysm that restoration of aortic con

tinuity is possible only by insertion of an arterial graft or a prosthetic tube. Creech[30] and Deterling[31] and Crawford and Debakey[32] have recommended prosthetic tubes of various plastic materials to bridge long arterial defects.

Technic of Excision and Anastomosis

The successful outcome of the contemplated operation begins with the preparation of the medial malleoli or into the right radial vein just proximal to the wrist These infusions are kept open until the operation begins with a slow drip of 5 per cent dextrose in water Thus, the anesthesiologist has ready access to the circulation for administration of intravenous anesthetic, transfusion, fluid, or medication

After the patient has been anesthetized and an endotrachial tube has been inserted, he is placed in the right lateral decubitus position (left chest up), his back along the edge of

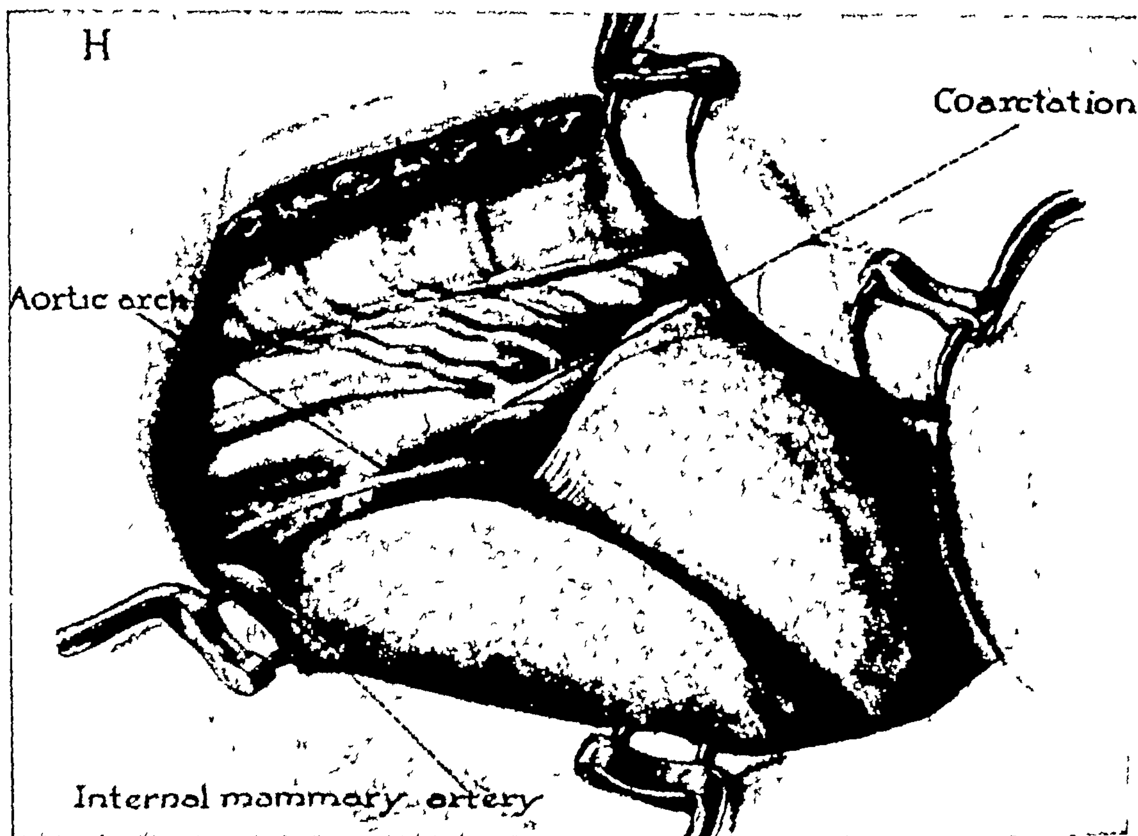


Fig 2 Pleural covered coarctation of the aorta as it appears upon opening the chest through the fourth intercostal space (Gross, R E Surgery of Infancy and Childhood W B Saunders Co, Philadelphia, 1953)

skin of the chest in the patient's room In this era of antibiotic resistant organisms emphasis is placed on the importance of the skin preparation The patient is shaved from mentum to umbilicus both front and back, including both axillae The shaved area is scrubbed for ten minutes with an antiseptic soap This is immediately followed by the application of sterile towels which are anchored in place with adhesive tape to prevent sliding On the day of operation a large plastic infusion cannula is inserted under local anesthesia into each of the saphenous veins just above the the table Once again the skin is prepared, after removal of the towels from the chest, by scrubbing again with an antiseptic soap The area is blotted dry with sterile towels and painted with an antiseptic solution after which the field is draped

A long posterolateral (parascapular) incision is made and carried down to the muscle layers Skin towels are applied In coarctation of Types I and IIA the muscular arteries are dilated and bleed furiously when divided It is good practice to lift up the muscular layer as they are divided, the operator making his

division in increments, meanwhile controlling the bleeding with the thumb and forefinger on one side of the cut muscle as an assistant does the same on the opposite side. Care should be exercised to preserve the continuity of the *internal mammary* artery and as many of the intercostals as is possible in order to maintain maximal collateral circulation while the aorta is clamped. Although electrocoagulation may be readily and quickly done should the need arise for wider exposure.

The lung is retracted manually with moist pads. The coarcted aortic segment (Fig 2) becomes readily visible under the glistening mediastinal pleura which is incised generously in a longitudinal direction. The vagus nerve and its recurrent laryngeal branch are identified and protected throughout the procedure.

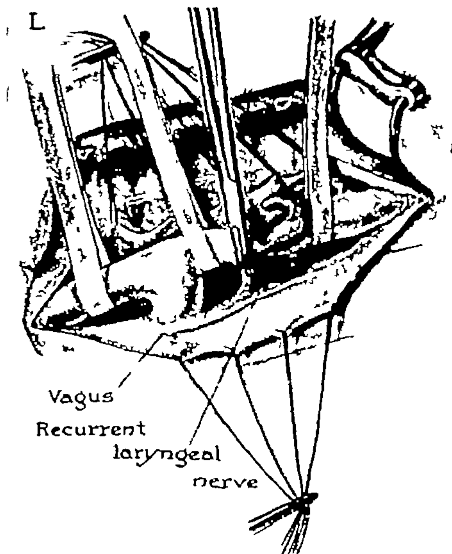


Fig. 3: Wide mobilization of the aorta by gentle sharp and blunt dissection aided by traction tapes. Note the right angle (Lahey) clamp applied to the ligamentum arteriosum. (Gross, R. E.: Surgery of Infancy and Childhood W. B. Saunders Co., Philadelphia, 1953.)

control smaller bleeding points and reduce the operative time the larger vessels should be properly secured with ligatures or suture ligatures. The incision is deepened and the thorax is entered via the left fourth intercostal space. After insertion of the rib spreader the ribs are spread gradually a little at a time in order to avoid rib fracture Ordinarily the ribs are not divided posteriorly but this can be readily and quickly done should the need arise for wider exposure.

Marsupializing sutures are applied to the cut pleural edges and are weighted with hemostats to assist in the exposure

The dissection is begun in the area of the aortic arch and left subclavian artery progressively *approaching* the stricture. As a safety measure and to enhance ease of dissection traction tapes may be passed about the subclavian artery and the proximal descending

aorta At the distal extremity of the pleural incision and between two adjacent pairs of intercostal arteries a tunnel is made beneath the distal aortic segment and another traction tape is passed to facilitate dissection (Fig 3) Each pair of intercostal arteries in the area of dissection is mobilized freely By this method or system of centripetal dissection the easier portion is completed first, leaving the more difficult and possibly hazardous part

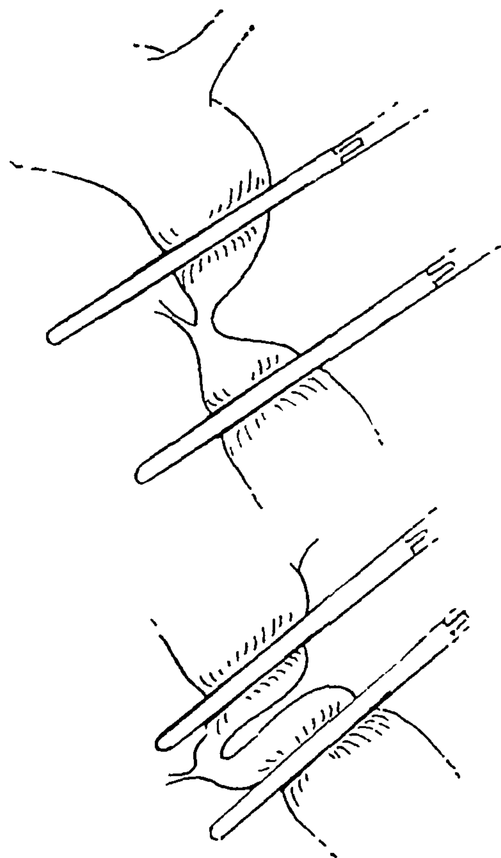


Fig 4 Upper—Application of occlusive clamps in desired position and at proper angle of inclination Lower—Determination of feasibility of reapproximation of aortic ends prior to division by tentative approximation of the clamps (Bailey, C P: Surgery of the Heart, Lea & Febiger, Philadelphia, 1955)

to the last Otherwise, should the operator get into technical difficulty early in the course of the aortic mobilization he is less able to deal with it or, possibly, even to complete the procedure The area most likely to produce difficulties is the region of the first two or three pairs of intercostal arteries immediately distal to the site of the narrowing

Great care and patience must be exercised in handling the dissection of intercostal vessels arising close to the coarctation for here they are most likely to be thin walled, perhaps even aneurysmal, and easily torn Even though it is an expressed purpose to conserve as much of the collateral circulation as possible, aneurysmal dilatation of an intercostal vessel requires its excision By steadfastly adhering to the principle of conservation of the existing collateral circulation damage to the spinal cord is precluded during the period of aortic clamping Furthermore, if for any reason the size of the lumen at the site of the anastomosis should be smaller than ideal a satisfactory result may still be achieved if all of the collateral passages are left intact

During the dissection of the ligamentum arteriosum (or patent ductus arteriosus) the recurrent laryngeal nerve is displaced medially Two clamps are applied across the ligamentum or ductus and it is divided between them If a lumen is present the pulmonary arterial end of the ductus is oversewn with 5-0 arterial silk, the aortic end being left clamped This clamp is removed later along with the coarcted segment (Fig 4)

Once the operator believes he has completed the dissection and is ready to divide the aorta he should inspect the field starting at that point at which he undertook the dissection With the aid of the traction tapes he should be able to elevate the aorta enough to see both the right and left sets of intercostal arteries Little strands of tissues left attached in the area of the anastomosis can lead to time consuming difficulty once suturing has begun It is essential that the field be dry and clean and that there be an immaculate dissection before proceeding to the final step It should be mentioned at this point that the adventitia should not be removed from the aorta for it is the strongest layer of the vessel and the one upon which the operator counts most for a blood tight anastomosis The conservation of this layer of tissue is especially valuable in securing leaks along the anastomotic line which may become apparent when the clamps are removed to reestablish the aortic flow

At this point in the operation the surgeon is able to visualize fully the extent and length of the involved segment and any poststenotic dilatation or aneurysm formation The optimal site for division above and below the lesion is decided upon by comparison of the calibres of the two segments especially with relation to the slope of their fusiform ends Any intercostal arteries which may have to be sacrificed or temporarily occluded with bulldog clamps are considered in view of the

aortic clamps which must be placed and the arterial anastomosis which is to be done. These intercostals should be taken care of at this time.

It is advisable to apply the clamps from the left side of the table (the operator's side) and at an angle of approximately thirty degrees from an imaginary vertical line. By so doing the surgeon will be able to rotate the clamps from time to time during the anastomosis if necessary to obtain exposure of the posterior aspect of the aorta. We prefer to use our own

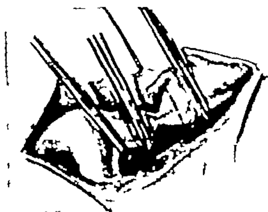


Fig. 5: The coarctation will be excised along the dotted lines. If the lesion were any closer to the subclavian artery a Satinsky clamp applied across both the aorta and the subclavian will usually allow sufficient cuff for anastomosis. (Gross, R. E.: The Surgery of Infancy and Childhood. W. B. Saunders Co., Philadelphia 1953.)

fenestrated toothed clamps applied at a sufficient distance from the aortic lines of incision to allow ample cuff for comfortable suturing. These clamps have unusually secure gripping characteristics which preclude slippage, are doubly fenestrated and have been described elsewhere[6, 31]. After the clamps have been closed, and before transection of the aorta, the feasibility of reapproximating the aortic ends after section is demonstrated by trying to bring the clamps into contact (Fig. 5). By this maneuver the elasticity of the aorta is assayed prior to excision of the lesion and before the surgeon has "burned his bridges." If the situation has been sized up correctly the lesion is excised making sure that the two aortic orifices are of the same size and of parallel obliquity (Fig. 6). By use of the clamps the aortic ends now may be approximated (making sure that there is no rotation of either end) and the anastomosis begun.

In children in whom continued bodily growth is expected it is theoretically best to employ an all-interrupted suture technic, using 5-0 arterial silk. However some surgeons use a continuous suture for the posterior row because of the rapidity with which it may be placed and because it is less likely to be accompanied by troublesome leaks. This aspect of the aorta may be approximated with

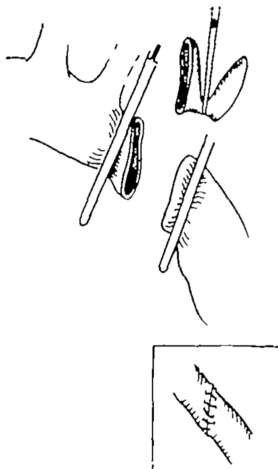


Fig. 6: Divided ends of the aorta are of equal size and obliquity. Such an anastomosis heals satisfactorily. (Bailey, C. P.: Surgery of the Heart. Lea & Febiger, Philadelphia, 1955.)

everting interrupted mattress sutures of 4-0 or 5-0 arterial silk or preferably by a continuous running mattress stitch (Fig. 7). Then the anterior aspect may be closed with either simple or mattress type everting interrupted sutures. In an adult patient future growth of the suture line is not to be considered. Therefore, both the posterior and the anterior row may be closed with a continuous running type stitch. From time to time during the period of suturing it may be beneficial to bathe the lumen with a dilute (1/1000) solution of heparin. As the final suture is tied down the

excluded portion of the aorta is filled with this solution to evacuate all air and to wash out any very small clots which may have formed

The next step is to ascertain the integrity of the anastomosis by filling the excluded segment with blood and this can be done by removing one of the serrefine clamps from an intercostal artery or by very slowly releasing the distal aortic clamp Clamps should be released slowly from the aorta, and the distal one should be removed first If bleeding occurs from a suture hole or from between two sutures it may be necessary to reapply the clamp to repair the leak If complete vascular integrity is not obtained immediately a longitudinally folded piece of dry gauze should be wrapped about the anastomosed area and held firmly in place for a period of five minutes or more after the distal clamp has been removed (Fig 8) The proximal clamp is loosened slightly during this time to allow some blood to traverse the anastomosis The anesthetist is alerted to check the blood pressure in the upper extremities constantly as the proximal aortic clamp is opened as sudden removal may produce a precipitous fall in blood pressure and even death[34] If the patient's condition remains satisfactory, a period of five minutes is spent slowly releasing this clamp, finally removing it If the blood pressure falls to lower than normal levels the pressure in the upper segment can be restored by reapplication of the clamp and, after a time, more cautiously loosening and removing it

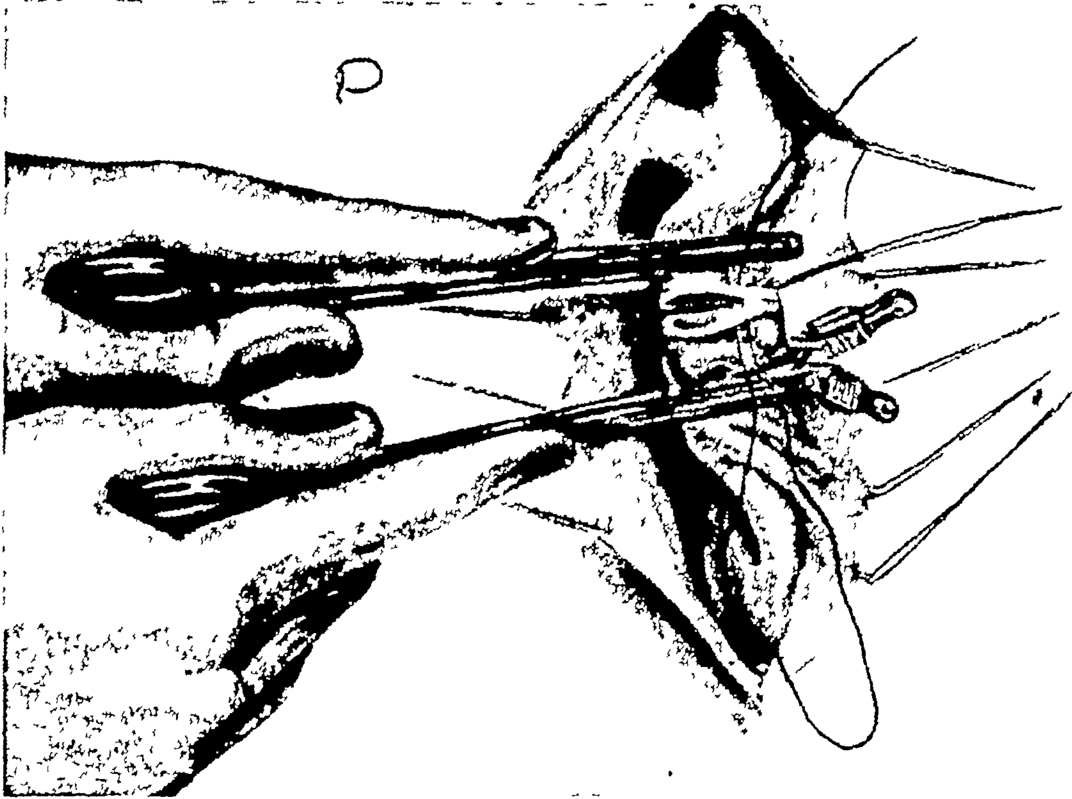


Fig 7 The clamps are rotated medially (toward the front of the patient) to expose the posterior aorta for suturing The aortic ends are approximated so that the suture line is under not the slightest tension (Gross, R E The Surgery of Infancy and Childhood W B Saunders Co, Philadelphia, 1953)

When one first applies the aortic clamps there may be a further rise in an already high blood pressure in the upper portion of the body perhaps to a very dangerous level Some operators have found it a safe and satisfactory policy purposely to allow blood replacement to lag approximately 500 cc behind actual loss When the clamps are released after completion of the anastomosis this unit of blood should be replaced to restore the blood volume to normal.

The incision in the mediastinal pleura is closed (after a final inspection of the suture line) leaving at its caudal aspect an opening for drainage A hemostatic search is carried out and then the chest is closed One or two

multiwindowed catheters of appropriate size may be inserted through intercostal stab wounds for postoperative (underwater seal) drainage of the pleural space.

Complications of Operation

Mortality: The operative mortality from the surgical repair of coarctation of the aorta was significantly higher during the development period than it is now. As improvements have been made in the medical management of infants seriously ill with coarctation to "tide them over" until a more opportune time for operation presents itself, better methods of case selection have been evolved, and as more experience has been gained by a trained group of surgeons doing the operation, a decline in the fatality rate has occurred. Since approximately one fourth of these patients have additional serious cardiovascular defects it is worthwhile to point out that the surgical risk in "pure coarctation" is very small. In our own hands it has been less than 2 per cent.[6] The risk of operation in individuals with one or more associated anomalies (excluding patent ductus arteriosus) probably is four to five times greater. In a collected series of 1601 operated patients reported by the American College of Chest Physicians in 1956 the mortality was 8.6 per cent.[35]

The operative risk is higher at the extremes of age. The management of the patient one year of age or less remains controversial. As mentioned, some internists believe prolonged medical treatment of these infants will yield a higher salvage rate while others feel operative intervention is indicated at the first suggestion that the baby is in difficulty. We believe that an infant with Type IIA coarctation (formerly called postductile) should undergo operation at the earliest opportunity. With the pulmonary congestion caused by the left-to-right shunt complications are likely to occur early and to be serious. These infants tend to go into congestive heart failure early in life and are subject later to the development of pulmonary arterial hypertension and arteriolar change. However, the operative mor

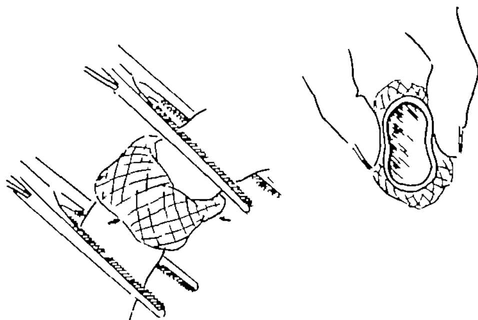


Fig. 8: A dry folded gauze encompasses the aorta and is very gently compressed during release of the aortic clamps. This practice aids in sealing the stitch holes and controls bleeding.

tality in group Type II B (infantile or preductile) lesions was found to be 28.7 per cent in the American College of Chest Physician's report[13] Probably this mortality was related to the lack of collateral circulation and might have been reduced by the use of hypothermia or a shunting bypass

With older patients in whom operation has for some reason been deferred too long one must be prepared to encounter complications such as cerebrovascular lesions, renal damage, myocardial insufficiency, aneurysm of the aorta or intercostal arteries, etc It becomes increasingly evident that the physician should urge these individuals to come to operation prior to the development of such changes A policy of postponement of surgical treatment past the teenage group may lead to catastrophe, elevates the operative mortality unnecessarily, and depresses unrealistically the statistical percentage of satisfactory results Cardiovascular failure is the leading cause of operative death in this group

One of the most disconcerting of operative complications is a paresis or paralysis of both lower extremities due to ischemic damage to the spinal cord during the period of aortic cross-clamping. Very frequently these subjects are adults which again points out the desirability of intervening earlier The employment of general bodily hypothermia or the use of a supplemental by-pass shunt (left atrium to femoral artery) with a pump will protect the spinal cord Patients with an underdeveloped collateral circulation are the ones most likely to develop this complication

While we have not encountered postoperative paradoxical arterial hypertension following resection of aortic coarctation such a complication is possible and may be fatal Sealy and associates[36] studied this problem and have offered several possible explanations for its appearance either early during the first 36 hours, at which time it is unimportant clinically or later when it is more ominous The delayed type of hypertension usually becomes manifest on the second postoperative day but has appeared for the first time as late as seven days after resection Severe abdominal pain often is associated with such paradoxical hypertension Usually the abdominal findings along with the hypertension diminish and disappear by the fourteenth to the twenty first day without sequelae Occasionally the pain may persist and merge into the signs and symptoms of necrosis of the small bowel which will necessitate resection

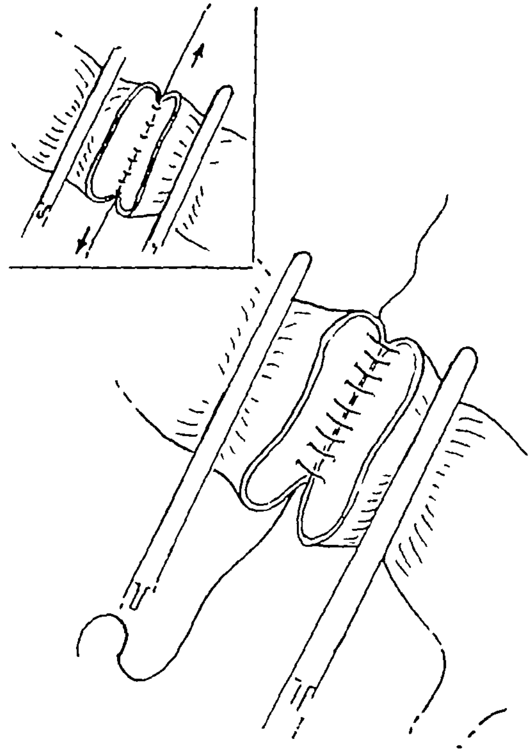


Fig 9 The continuous over and over suture has been shown to be the preferred technic in the hands of the average surgeon (Bailey, C P Surgery of the Heart, Lea & Febiger, Philadelphia, 1955)

The responsible arterial lesions involve the smaller arteries which arise distal to the resected portion of the

aorta, usually those to the small intestine. Pathologically the smaller arteries and arterioles present the appearance of a necrotizing panarteritis the lesions being histologically similar to those of periarteritis nodosa and closely resembling those seen in experimental hypertension.[36]

Disruption of the aortic anastomosis is an important mechanism of death in surgery for coarctation. The usual cause of the disruption is infection at the site of anastomosis. About half of these fatalities are due to localized infection about the line of anastomosis. Empyema, septicemia, or subacute bacterial endocarditis contribute to the rest. In addition to the routine use of pre and postoperative antibiotic therapy in an attempt to prevent these complications an effort should be made to keep to a minimum the period of time the chest is open and its contents exposed to the possibility of airborne infection.

Some surgeons believe the suture technic used in the anastomosis may have a direct bearing upon the incidence of disruption. A simple continuous noneverting suture technic has been associated with less than half the fatality rate reported with employment of the everting mattress suture method[14] (Fig 9) The higher incidence of disruption in the latter group might be related to imperfect placement and tying the sutures tightly enough to cause pressure necrosis. These observations suggest that the mattress technic especially in interrupted form should be employed only by unusually capable operators. An interrupted simple suture technic permits unrestricted growth of the anastomosis and is preferable for infants. In larger children the posterior or less accessible aspect of the anastomosis may be performed with a continuous running suture while the presenting aspect may be closed with interrupted stitches.

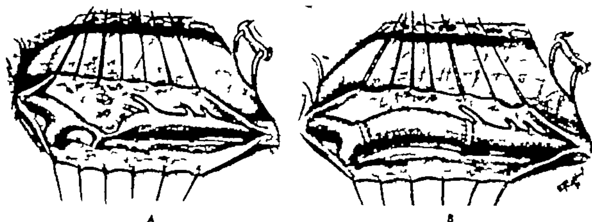


Fig 10: A shows a long segment of aorta which must be excised leaving a gap too long for end to end anastomosis of the cut aortic ends. B shows the long constricted area replaced by a graft (Gross, R. E.: The Surgery of Infancy and Childhood W. B. Saunders Co Philadelphia, 1953.)

The danger of uncontrollable hemorrhage from thin walled, friable, and dilated intercostal arteries has been mentioned The aortic ends, especially in adults with very long standing disease likewise may be thin walled and present similar dangers This reemphasizes the desirability of earlier operation before degeneration has occurred. The incidence of fatal bleeding associated with insertion of a homologous arterial graft is understandably higher than that with end-to-end anastomosis of the aortic ends (Fig 10) The incidence of

aneurysm formation following aortic resection and replacement is uncertain because of the relatively short period of follow-up in terms of years Our experience with experimental operations suggests that the rate of aneurysm development will be higher with arterial homografts than with prosthetic tubes There are several theoretical reasons for this There is essentially no immunologic reaction to the inert synthetic material Both the homograft and the prosthesis act primarily as scaffolds about and upon which scar tissue is formed With the preferred plastics the strength of the synthetic remains at a relatively constant level while the tissue of the homologous artery becomes absorbed with the passage of time[32] Once an aneurysm has developed immediate resection is indicated Often a prosthetic tube will be required to bridge the resulting defect

Exploratory Operation

An exploratory operation for the purpose of establishing the diagnosis or resectability of coarctation is hardly justifiable Modern radiographic technics may be employed in equivocal cases to delineate the extent, type of obstruction, and aneurysmal formation if present This information will prepare the surgeon for the probable necessity for use of a graft or prosthesis at the time of operation With the general availability of preserved arterial homografts and prosthetic tubes and with the progressively increasing availability of cardiopulmonary by-pass technics there is less and less justification for a nondefinitive exploratory operation

Results of Operation

In general it may be stated that coarctation of the aorta is one of the most gratifying cardiovascular lesions to treat surgically The results in terms of satisfactory relief of hypertension exceed 90 per cent in most series, more than 70 per cent showing a return to completely normal blood pressure relationships in the upper and lower extremities, another 20 per cent obtaining marked lowering of the upper extremity blood pressure In a small percentage the arterial hypertension persists, perhaps because of some undiagnosed associated lesion

Patients who have a satisfactory technical result but still manifest upper extremity hypertension should be submitted to a complete hypertensive work-up Some of these patients will be found to have a surgically remediable disease Certain of these subjects should be kept under antihypertensive medical management

It should be emphasized that in nearly every instance of surgery for coarctation with reestablishment of an adequate aortic passageway the patient's general health will be improved By removal of the arterial blockage the status of the cardiovascular and renal systems is benefited significantly

Usually the blood pressure falls appreciably within days of operation but it may require several weeks to descend to completely normotensive levels Should the arterial pressure remain elevated for several months the patient is not likely to receive further benefit from the operation and arrangements should be made to investigate the etiology of the persisting hypertension.

In the very young and in older patients the risk is greater and the results less striking than in the mean ages although approximately 96 per cent of the patients who survive surgery are improved

References

1 Gross, R. E. The Surgery of Infancy and Childhood, W B Saunders Co., Philadelphia, 1953
2. Bonnet, L. M. Rev de med Paris, 23 108 255 335 418, 481 1903
3 Edwards, J E. Malformation of the Aortic Arch System in Pathology of the Heart, S. E. Gould. Charles C. Thomas, Springfield, 1953
4 Bailey C. P. Surgery of the Heart, Lea & Febiger Philadelphia, 1955
5 Lemmon, W M and Bailey C. P J Thoracic Surg 35 391 1958.
6. Bailey C. P., Lemmon, W M and Musser B. G. "Experiences with Coarctation of the Aorta" (To be published American Journal Cardiology)
7 Martin, J F and Yount, E. H Am. J Roentgenol., 76 782, 1956.
8 Bailey C. P Surgery of the Heart, Lea & Febiger Philadelphia, 1955 p 137
9 Campbell, M. and Baylis, J H Brit. Heart J., 18 475 1956.
10. Abbott, M E. Am Heart J 3 574 1928.
11 Reifenstein, G H., Levine, S. A., and Gross, R. E. Ibid., 33 146, 1947
12. Inmon, T W and Pollock, B S. Ibid., 52 314 1956.
13 Bailey C. P., et al Dis. Chest, 31 468, 1957
14 Shanahan, W R., Romney S L., and Currens, J H J.A.M.A 167 275 1958.
15 Sandifer S H Am. Heart J., 51 761 1956.
16. Gerbode, F., et al. A.M.A. Arch. Surg 72 850, 1956
17 Neuhauser E. B D and Wittenborg, M H Circulation, 11 471 1955
18 Lemmon, W M and Lehman, J S. "Transthoracic Aortography" (Unpublished data)
19 Mustard, W T., Rowe, R. D., Keith, J D., and Sirek, A. Ann. Surg 141 429 1955
20. Lang, H T., Jr and Nadas, A. S Pediatrics, 17 45 1956.
21 Johnson, J and Kirby C. K. Surgery 27 17 1950.
22. Blades, B and Pierpont, H C. Am Surgeon, 21 739 1955
23 Alexander J and Byron, F X. J.A.M.A., 126 1139 1944
24 Blalock, A. and Park, E. A. Ann. Surg., 119 445 1944
25 Clagett, O T. Proc. Staff Meet. Mayo Clin., 22 131 1947
26. Johnson, Jr., Kirby C. K., and Hardy J A Surgery 33 207 1953
27 Wiggers, C. J Circulatory Dynamics, Grune and Stratton, New York, 1952, p 70.
28. Holman, E. J Thoracic Surg., 28 109 1954
29 Gross, R. E. and Meeker L. A., Jr Surgery 29·419 1951
30. Creech, O Jr., et al. Ibid 41 62, 1957
31 Deterling, R. A., Jr and Bronshay S. B Ibid., 38 71 1955
32 Crawford, E. S DeBakey M E., and Cooley D A. A.M.A. Arch. Surg 76 261, 1958.
33 Bailey C. P and Musser B C. J Thoracic Surg 35 549 1958.
34 Gross, R. E. and Hufnagel, C. A. New England J Med., 223 287 1945
35 Rumel, W R., et al. J.A.M.A., 164 5 1957
36. Sealy W C., Harris, J S Young, G., and Callaway H A. Surgery 42 135 1957

VASCULAR COMPRESSION OF THE ESOPHAGUS AND/OR TRACHEA

WILLIAM M LEMMON, M D., AND CHARLES P BAILEY, M D

Anomalies of the great vascular structures of the thorax probably are more common than is generally appreciated for many of these malformations are compatible with long life and freedom from symptoms Recognition and treatment early in life of those abnormalities of the great vessels of the thorax which produce significant compression of the esophagus and/or trachea is essential and may be life saving The clinical picture presented by these subjects along with rather minimal investigative studies usually will lead to sufficient diagnosis of the anomaly to permit surgical alleviation to be carried out in time

Vascular malformations of the aortic arch and its branches assume many diverse forms and may be very complex. While Hommel (1737)[1] published an illustration of a double aortic arch and Bayford (1794)[2] described dysphagia from esophageal impingement due to an anomalous right subclavian artery it was not until 1939 that Wolman[3] described the clinical syndrome caused by the double aortic arch. Arkin (1926)[4] described the roentgenographic aberrations of the aortic arch and these were further elaborated by Neuhauser[5] However, it has been due largely to the writings and teachings of Gross[6] who first reported successful surgical management of these anomalies that proper attention and focus has been brought to bear upon this subject.

If the diagnosis is suspected radiographic investigation of the esophagus and/or trachea employing iodized oily or aqueous solution is indicated This must include not only posteroanterior but lateral and oblique roentgenograms as well It is dangerous to use contrast mixtures containing barium in these small patients because of the likelihood of spillage of material into the tracheobronchial tree with resultant severe pneumonitis.[7] Aqueous iodized solutions disappear from the bronchial tree much more quickly than oily preparations Once the diagnosis of compression of the trachea or esophagus, or both has been established by radiographic means, the exact anatomic diagnosis may depend upon more elaborate studies such as angiocardiography or aortography These special procedures entail a certain amount of risk and should be avoided whenever feasible.

In the future additional types of malformations may be recognized and become amenable to surgical treatment. At present five different abnormalities are described generally all of which are partially or totally correctable by operation. The anatomic variants under discussion include (1) double aortic arch, (2) right aortic arch with a posterior ligamentum arteriosum, (3) anomalous innominate artery (4) anomalous left common carotid artery and (5) anomalous right subclavian artery

Double Aortic Arch

Pathology: The normal human aortic arch passes anterior to the trachea, proceeds to the left, deviates dorsally and then turns caudally to become the descending thoracic aorta. Persistence of both fourth branchial arterial arches during intrauterine life results in a double aortic arch. This is the most frequently encountered of this group of

comparatively rare lesions of the aortic arch system (Fig. 1). The anterior limb of the double aortic arch is the successor of the left fourth primitive arch while the posterior limb represents the

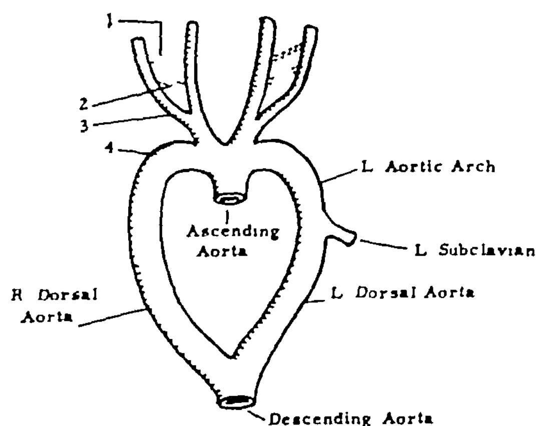


Fig 1 Diagram of persistence of both fourth branchial arterial arches producing double aortic arch

right arch Thus the ascending aorta in such instances bifurcates into two branches, one passing in front of the trachea and the other posterior to the esophagus to join again with its opposite member to resume continuity as the descending thoracic portion of the aorta The ligamentum arteriosum (obliterated ductus arteriosus) joins the aorta just distal to the site of reunion of the arterial segments In most instances the anterior or left arch is the smaller but in some cases the posterior arch is the smaller of the two Usually the posterior branch gives rise to the innominate artery while the left common carotid and left subclavian arteries more commonly arise from the anterior arch (Figs 2 and 3) Rarely the anomaly is associated with a right descending aortic arch, the descending aorta then lying to the right of the vertebral column

Symptomatology: Although but minor degrees of compression of the trachea and/or esophagus may result from a double aortic arch which then is compatible with a long life free of symptoms, commonly the anomaly becomes symptomatic during the first year or two of life Because of the small size of the lumen of the trachea in infancy, even a moderate amount of external compression may produce impressive respiratory symptoms Encroachment upon the esophagus may produce swal-

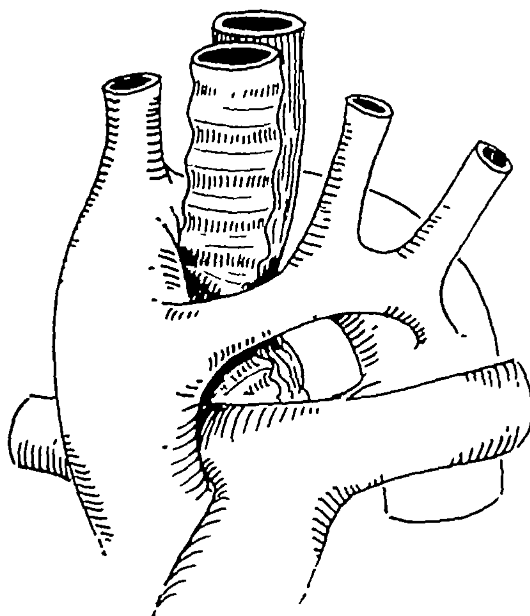


Fig 2 Anteroposterior view of a double aortic arch Note smaller anterior (left) arch and larger posterior (right) arch The smaller arch gives origin to the left bracheocephalic arteries This is the most frequent form of the anomaly

lowing difficulty but this usually is overshadowed by the respiratory distress Failure to bring the baby promptly to corrective surgery frequently will lead to fatality

Any infant presenting symptoms referable to breathing or swallowing should be suspected of having some type of vascular anomaly along with other possible malformations within the thorax Double aortic arch may cause intermittent or chronic symptoms related either to respiration or to the ingestion of food Usually the chronic pattern predominates Often the patient presents an alarming increase in

the rate of breathing with inspiratory retraction of the soft tissues in the abdominal, the intercostal and the suprasternal and supraclavicular areas. Upon auscultation of the chest there will be heard a loud inspiratory and expiratory wheeze which may be audible even

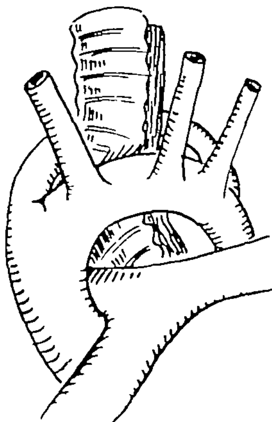


Fig. 3 Vascular pattern sometimes seen showing the larger anterior arch giving origin to the innominate artery. The left common carotid and left subclavian arteries also arise from it.

when standing at the bedside (crowing type of respiration) In addition to the stridor there will be coughing and often associated episodes of choking and cyanosis especially following ingestion of milk or food. The position assumed by these small patients is almost pathognomonic of the condition. It is one by which they can improve the lumen of the partially obstructed airway. They tend to assume the prone position and hold the back and head in marked hyperextension. If there is superimposed tracheobronchitis or pneumonitis the impression of subepiglottic obstruction will persist even after all mucus has been aspirated from the tracheobronchial tree. In spite of immediate intensive medical therapy these babies emerge from the acute episodes with a persistent picture of chronic respiratory obstruction.

Radiologic Findings: The most important definitive examination is an adequate roentgenologic study which

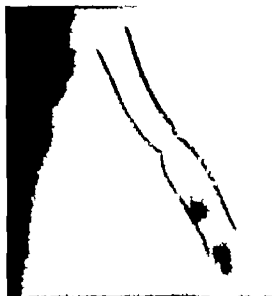


Fig. 4 Retouched lateral roentgenogram showing area of compression due to double aortic arch as outlined by air filled trachea

includes outlining of the esophagus and/or trachea with a thin coating of contrast material. Routine chest films may show evidences of pneumonia and these same views may reveal air in the trachea which helps to outline the site of compression. The lateral view frequently is most helpful in this respect (Fig. 4). The lower trachea is pressed forward with marked reduction in its lumen by a smooth external structure. Contrast outlining of the esophagus reveals an indentation of the posterior esophageal wall at the level of the third or fourth thoracic vertebra (Fig. 5). A tracheogram (often an inadvertent associated study) will demonstrate a

constriction upon both sides of this air passage in the oblique view but the lateral view usually reveals merely an anterior indentation in its lower third (Fig 6) Radiologic evidence of external impingement upon the lumina of both the trachea and the esophagus at approximately the same level is pathognomonic of vascular compression From a practical viewpoint, aortography is not recommended in these cases even though it may outline the pathologic anatomy precisely (Fig 7) This additional special study, with the inherent risks which it imposes upon an already seriously ill infant, does not influence the course of therapy materially and generally should be avoided



Fig 5 Posteroanterior view of contrast study of esophagus showing defect on the posterior wall due to double aortic arch



Fig 6: Oblique roentgenogram (with spillage of contrast material from the esophagus into the trachea) illustrating narrowing on both sides of the trachea due to double aortic arch



Fig 7 Superimposed diagram of trachea upon aortogram demonstrating double aortic arch

Surgical Treatment: Since the surgeon will not know prior to operation which of the two aortic arches will have to be divided, it is desirable to perform a long lateral incision through the left fourth intercostal interspace It is essential that all of the large mediastinal structures and all branches of the respective aortic arches be mobilized in order to ascertain the exact anatomical variant involved The usually rather large thymus gland may be sacrificed by removal, or it simply may be retracted from the field of dissection Both

of the arches must be visualized clearly for the smaller is to be selected for division. In the vast majority of patients who have a left descending thoracic aorta the left or anterior arch will be the smaller and will be divided electively. In a significant number of cases there is a right descending aorta (Fig 8). Even in this group usually the left or anterior arch is smaller and, therefore, should be divided (Fig 9A), along with the ligamentum arteriosum and perhaps the subclavian artery (Fig 9B).

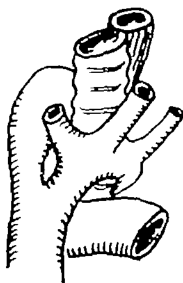


Fig 8: In this case of double aortic arch (front view) there is a right descending aorta. The left (anterior) arch usually is the smaller one in such instances and then is the one to be divided.

The left common carotid artery frequently arises from the smaller anterior arch and it usually is preferred technic to divide this arch distal to the carotid origin, *i e* between it and the subclavian artery (Fig 10). This latter artery may be sacrificed if any purpose is served thereby but an attempt should be made to preserve it as the primary source of arterial blood to the left upper extremity. Usually however no ill effects result from ligation. If after dividing and oversewing the cut ends of the anterior arch the left common carotid artery continues to lie upon the trachea the vessel should be drawn forward (ventrally) (Fig 9B) and tacked to the back of the sternum with several heavy mattress sutures which pass through the arterial adventitia and the sternal substance thereby eliminating any tendency toward perpetuation of the compression of the airway.

Early in the dissection the ligamen

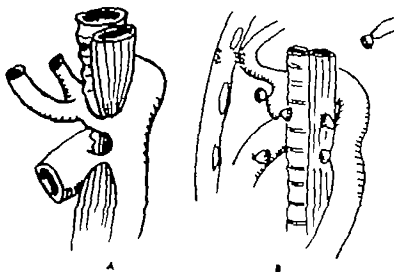


Fig. 9: A, The right aortic arch of this double aortic arch (lateral view) is to be preserved while the smaller left (anterior) arch should be divided close to its junction with the descending aorta. B The ligamentum arteriosum and the first part of the left subclavian artery also have been divided while the left common carotid has been drawn away from the trachea by suturing to the sternum.

tum arteriosum should be divided to allow the pulmonary artery to fall away from the systemic arteries thus facilitating the remainder of the dissection as well as further releasing the enveloped and perhaps restricted structures Care should be exercised throughout the operation to avoid undue trauma or frank injury to the recurrent laryngeal or the phrenic nerves

If after completely freeing up and visualizing all structures the posterior arch is found to be the smaller, it will be the one selected for division (Fig 11 A,B,C) Usually it is most convenient to place two vascular clamps across the vessel immediately proximal to the point of junction with the descending aorta Either the Potts multi-toothed clamps or the Bailey-Musser fenestrated clamps may be used If there are no detectable changes in the pulses of the head, neck, and upper extremities following application of these clamps and if the vital signs remain stable after the passage of several minutes, it is safe to divide the artery between clamps and to oversew the ends When the fenestrated clamps are used two parallel lines of mattress sutures may be placed across the vessel before severing it At this point or previously one should

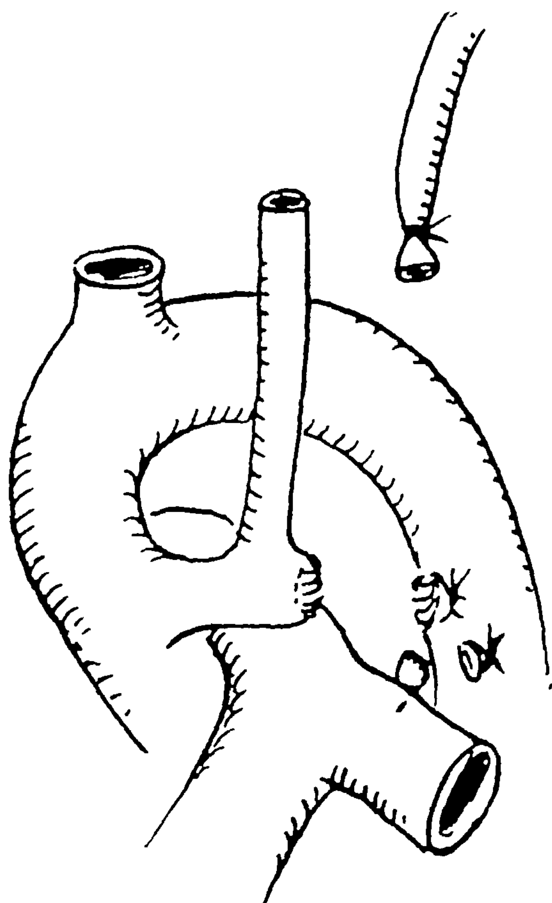


Fig 10 Postoperative situation in the most frequently encountered anomaly The smaller anterior or left arch has been divided just distal to the take off of the left common carotid artery and the ligamentum arteriosum has been divided The left common carotid should be sutured to the under surface of the sternum to avoid possible tracheal pressure if necessary Often the left subclavian artery will be divided to further mobilize the structures

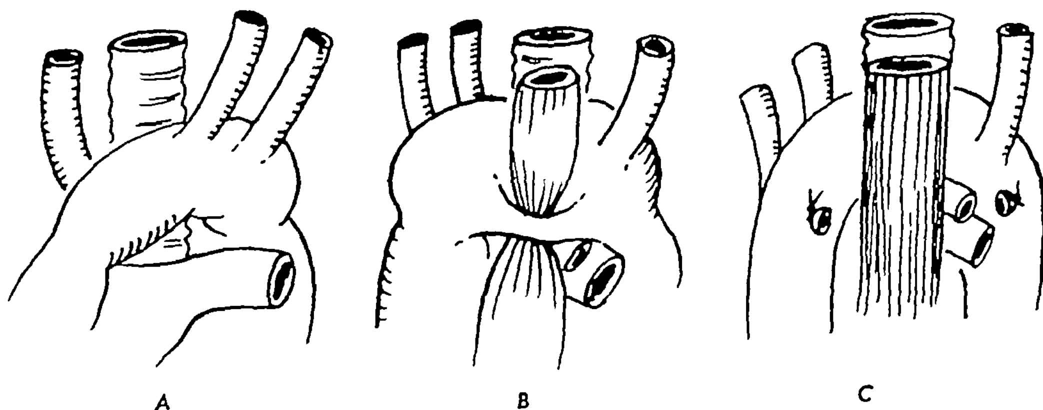


Fig 11 A case of double aortic arch in which the posterior arch is the smaller *A*, Anterior view prior to division of the constricting bands *B*, Posterior view of *this rare anomaly* *C*, The view from the posterior aspect after division of the smaller arch

of double aortic arch to be under six months while most of those with right aortic arch and constricting ligamentum arteriosum were several years older. The clinical picture is not dissimilar to that seen in patients with double aortic arch but tends to be less severe Physical findings include inspiratory retraction of the suprasternal and supraclavicular regions, overactivity of the accessory respiratory muscles, stridor, crowing respiration, occasionally dysphagia, and perhaps evidence of pneumonitis

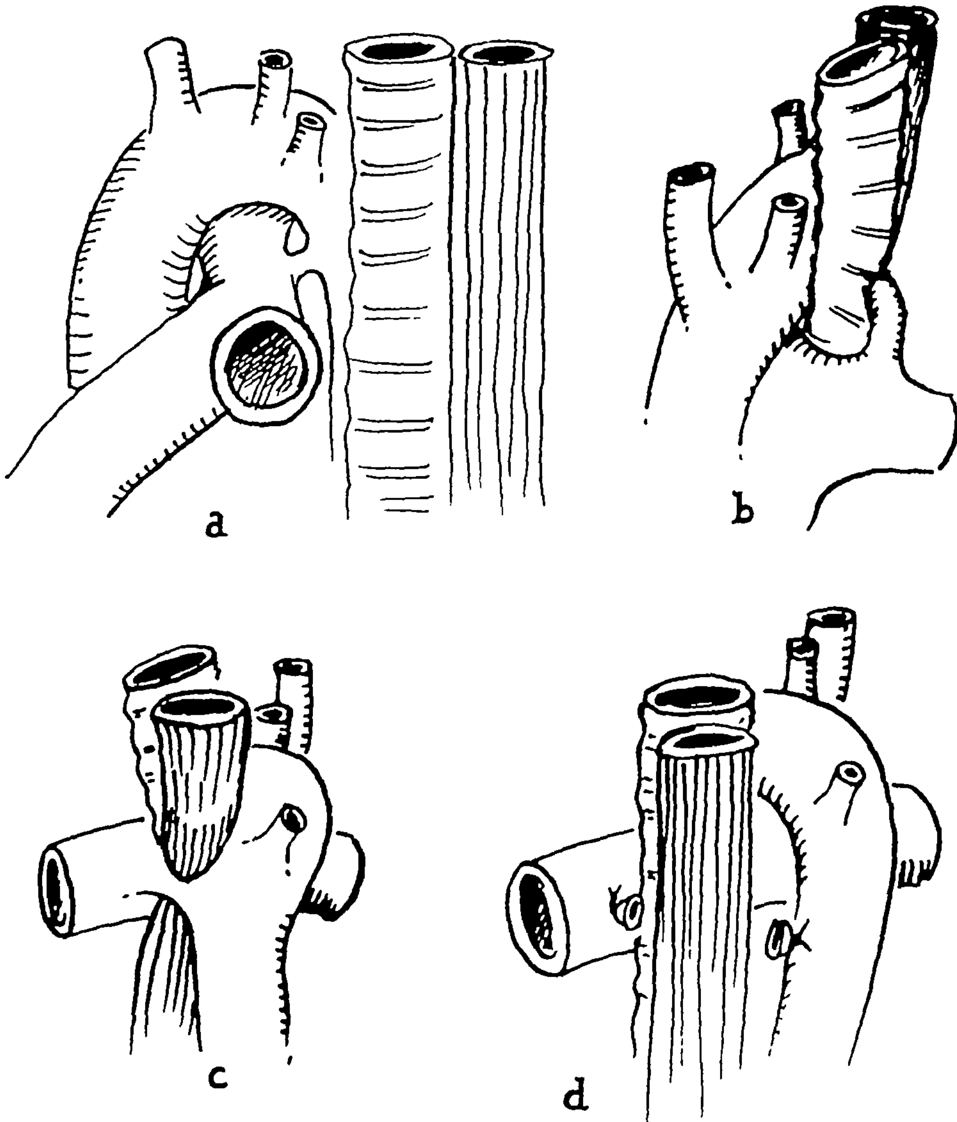


Fig 13 *a,* Nonconstricting right anterior ligamentum arteriosum with right aortic arch (left lateral view) *b,* Right aortic arch with right posterior ligamentum arteriosum compressing trachea and esophagus from the left (front view) *c,* Posterior view *d,* Posterior view after division of the ligamentum arteriosum

Radiologic Findings: Radiographic films of the chest taken during an acute symptomatic episode may reveal pneumonitis associated with a prominent right aortic knob. If the films are of high quality narrowing of the lower segment of the trachea may be seen with expiratory emphysema and underaeration of the lung fields during inspiration In the anteroposterior tracheogram performed with lipidol or one of the modern aqueous iodinated contrast media there usually will be noted on the right side of the outlined

structure a comma shaped, prolonged indentation caused by pressure from the right aortic arch. *In the lateral view* the ligamentum arteriosum may be seen to impinge upon the trachea Often it will "pull the pulmonary artery onto *the anterior* tracheal surface thus altering its outline at this site also. Contrast delineation of the esophagus will demonstrate a similar external pressure point at the same level upon its posterior and left lateral surfaces. There also may be evidence of impingement of the left subclavian artery against the posterior esophageal wall at a slightly higher level as it arises from the posterior arch and crosses obliquely behind the gullet to reach its exit from the thorax. Aortography usually is not indicated for it contributes nothing toward the surgical management of these cases and imposes an additional hazard.

Surgical Treatment: At the time of thoracic exploration it is essential for the operator to obtain wide exposure of the structures of the mediastinum, especially *posteriorly (Fig 13C) All* major vessels should be liberally mobilized and identified, as should the nerves. The constricting ligamentum arteriosum *should* be divided (Fig 13D) If it should be patent the cut vascular ends must be oversewn Finger dissection in the area of the lesion may be employed to make certain that all fibrous bands are cleared from the trachea and esophagus thus obviating the possibility of persistence of any obstruction from this secondary source. If the left subclavian artery is found to impinge upon the esophagus posteriorly it should be divided at its origin, its two cut ends

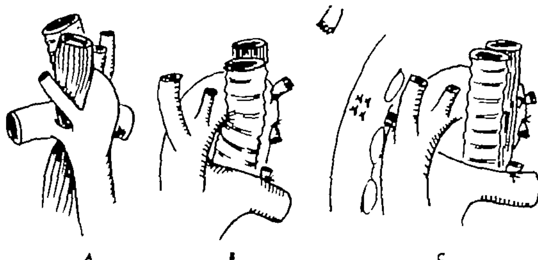


Fig 14 A, Left posterior view of the anomaly (right aortic arch with posterior ligamentum arteriosum) with a left subclavian artery impinging upon the esophagus. B, Surgical treatment of the malformation (anterior view) Division and oversewing of divided ends of ligamentum arteriosum and the left subclavian artery C The arch is sutured to the sternum to alleviate any tend ncy toward residual compression of trachea.

being oversewn with arterial silk (Fig 14A,B) This step insures the maximum possible clinical relief from encroachment *upon the encircled or involved* structures. Obviously a right aortic arch cannot be returned to the more usual anterior position. As a final step in the operative treatment the aorta and perhaps the pulmonary artery should be "tacked" away from the mobilized trachea and/or esophagus by placing several heavy braided silk sutures through the arterial adventitia and the soft bone of the sternum and tying

them with sufficient tension to pull these great vessels away from the trachea (Fig 14C)

Results of Surgery: The beneficial effect of surgery in this group of patients is gratifying for the mortality is almost nil, the morbidity is small, and the relief of symptoms usually is both prompt and striking There is disappearance of dysphagia and stridor, absence of retraction of the supraclavicular spaces, and great improvement in the general well being of the child

Anomalous Innominate Artery

Pathology: The syndrome of vascular compression of the trachea can be produced by aberrations in the origin and course of the innominate artery In this condition the pathologic anatomy usually assumes one of two readily recognized forms The more frequent of the two consists of a common origin of the innominate and left common carotid arteries at a point farther along the aortic arch than is usual (Fig 15A) In this circumstance the innominate artery must pass anterior and to the right of the trachea in its course to the right side of the thoracic inlet The severity of the symptoms produced depends upon the tautness with which the vessel encroaches upon the anterior wall of the trachea Minor degrees of the anomaly are fully compatible with a normal symptom-free life In the other type of case the anomalous origin of the (independent) innominate artery occurs at a more distal site along the arch than is normal (Fig 15B)

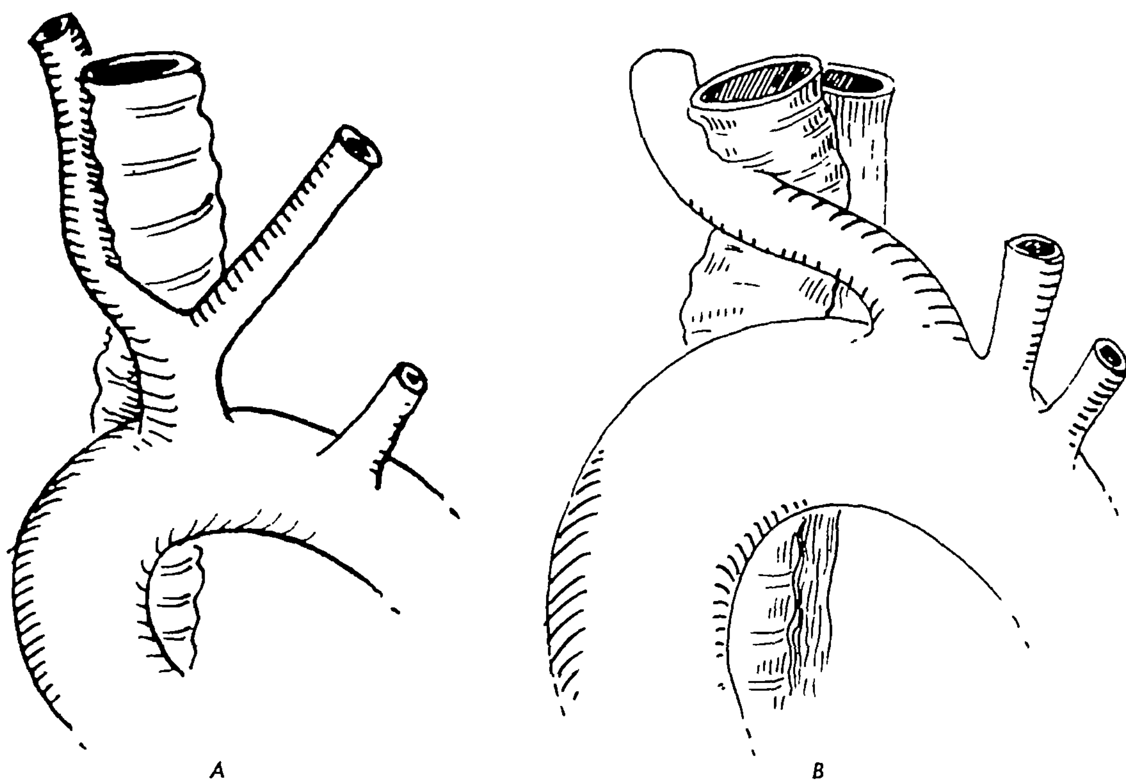


Fig 15 A, Pathology of anomalous origin of innominate artery in common with left carotid artery This produces an oblique line of pressure upon the trachea B, The innominate artery in this case arises independently and more to the patient's left than is normal In reaching the right thoracic outlet it compresses the anterior wall of the trachea in oblique fashion

Symptomatology: The signs and symptoms resulting from this lesion are similar to those described in the previous conditions and are likely to

develop early in life if the degree of compression is of an important grade. Differentiation cannot be made upon history or physical examination but the lesions can be distinguished with the aid of contrast studies of the trachea.

Radiologic Findings: Routine posteroanterior films of the thorax may show nothing unusual but the lateral view may show an anterior diagonal line of narrowing of the air filled trachea. Esophagography will reveal no abnormal finding while the tracheogram demonstrates the vascular compression of the anterior surface of the trachea and a normal posterior tracheal wall. In a given case it may not be possible to determine whether the long indentation of the trachea is due to innominate or left common carotid artery compression. Surgically this differentiation is unimportant. It is more important to determine if the lesion is due to tracheal malacia (faulty development of the cartilagenous rings of the trachea). Serial films made at different phases during the respiratory cycle are capable of differentiating the two conditions. If the trachea is compressed by a vascular component the indentation will remain present on all observations while if it is due to tracheal malacia it will be observed only during the expiratory phase of respiration.

Surgical Treatment: After entering the chest through a long lateral intercostal incision suitable exposure of the aortic arch may be obtained by reflecting the thymus and mediastinal pleura. There may be a common origin of the innominate and left common carotid arteries at a point farther along the aortic arch (toward the patient's left) than is usual or the innominate may arise independently at such a site. The artery passes to the right crossing anterior (ventrad) to the trachea compressing it (Fig 15A,B). The tracheal compression can be reduced by placement of a number of heavy mattress sutures of silk through the adventitia of the innominate artery (taking care not to pierce the lumen) and through the sternum. They should be placed extensively along the course of the vessel so that the total area of encroachment is relieved (from base to top of the indentation of the trachea). The sutures once placed should be pulled up together so that an even distribution of traction is exerted upon the arterial adventitia (Fig 16).

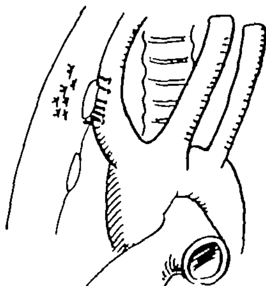


Fig 16: Surgical relief of tracheal compression due to anomalous origin of innominate artery by placement (and tying) of a line of heavy mattress sutures through the arterial adventitia and the sternum. The sutures are pulled up simultaneously to avoid pulling out or tearing the adventitia. Thus maximum relief from compression of trachea is provided. The same principles of treatment apply both in common origin of the innominate artery with the left carotid and in anomalous innominate artery originating independently from the aorta but far to the left.

Results of Surgery: This is a very satisfying lesion to treat, from the surgeon's standpoint, for the risk of operation is very low and the clinical results are excellent. The morbidity depends upon the quality of the surgical care postoperatively.

Anomalous Left Common Carotid Artery

Pathology: A common origin of the innominate and the left common carotid arteries may be located at a more proximal site along the aortic arch than is normal In such a case the left carotid arises more to the patient's right than usual and must pass in a diagonal direction across the front of the trachea to reach the left side of the inlet of the thorax In its course it may impinge upon the trachea deforming it along an oblique line Less commonly the left common carotid artery may originate independently from the aortic arch but far to the right Again it will follow a course similar to that described to produce an oblique line of compression upon the trachea (Fig. 17).

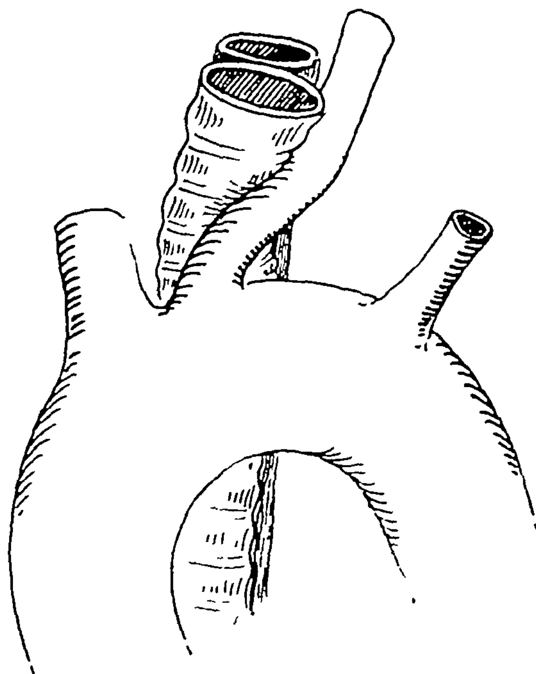


Fig 17 Left common carotid artery arising to the right of its normal origin from the aortic arch producing a diagonal line of compression of the trachea upon its left anterolateral aspect

Symptomatology: The more severe the degree of encroachment upon the trachea the earlier in life will symptoms appear. This anomaly provides one of the less common forms of compression of the trachea, and one in which the esophagus is not involved The clinical aspects of the malformation may be similar to those encountered in double aortic arch The patient may present a history of repeated bouts of respiratory infection since birth with intercurrent episodes of breathing difficulty such as dyspnea, crowing respiration, and utilization of the accessory respiratory muscles

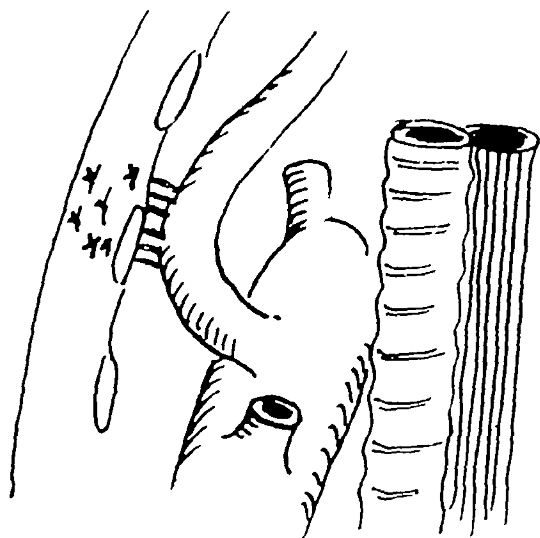


Fig 18 The tracheal compression is relieved by tacking the anomalous left common carotid artery ventrally by passing several heavy mattress sutures through the arterial adventitia and through the soft sternum.

Radiologic Findings: The observations made at this examination are similar to those made in examination of a patient with anomalous innominate artery except that the anterior tracheal defect follows a course diagonally upward from right to left as it travels toward the left thoracic outlet

Surgical Treatment: The clinical similarity of this condition to anomalous innominate artery extends to the surgical treatment as well. This malformation likewise responds to surgical tacking of the vessel to the posterior

aspect of the sternum by the passage of nonabsorbable sutures through the arterial adventitia and through the sternal substance, thus drawing the vessel ventrally so that the compression upon the trachea is relieved (Fig 18)

Results of Surgical Treatment: The mortality and morbidity from operative treatment of this anomaly are extremely low and the surgical results are quite satisfactory

ally it passes between the trachea and the esophagus as in Bayford s case, but more commonly it passes posterior to *both the trachea and esophagus.* Therefore, in its ascent from the left side of the aortic arch to the right side of the thoracic outlet it may produce esophageal compression and swallowing symptoms while the trachea characteristically remains free of involvement (Fig 19A,B)

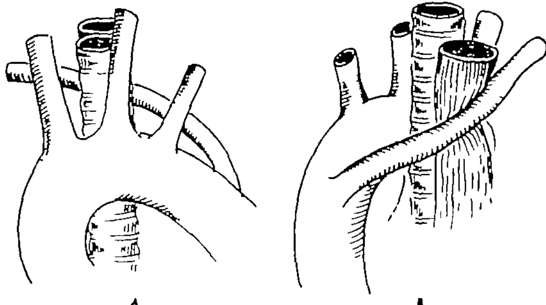


Fig. 19: A, Anterior view of anomalous right subclavian artery B Posterior view of the aberrant artery showing diagonal compression of the esophagus.

Aberrant Origin of the Right Subclavian Artery

Pathology: Since the description of "dysphagia lusoria by Bayford[2] in 1794 the swallowing difficulties which may be associated with an anomalous origin of the right subclavian artery have been known. It is not an uncommon finding to observe the right subclavian artery originating independently from the aortic arch. When it arises from the distal portion of the arch it must pass the trachea and esophagus to reach its normal anatomical position at the right thoracic outlet. Rarely does the artery pass in front of the trachea. Occasion

Symptomatology: Many individuals with this anomaly remain asymptomatic throughout life, the presence of the lesion being recognized merely by an incidental observation of a radiologist *during upper gastrointestinal examination.* The severity of the clinical picture varies greatly and is considered to be related directly to the tightness of the aberrant vessel and the degree of obstruction of the esophagus produced thereby In an infant there may be regurgitation of food after a few mouthfuls with immediate resumption of suckling for it is obviously hungry and unsatisfied. Some babies may be able to tolerate a very small amount of feeding

at a time and the mother may learn by experience to feed it more frequently and in smaller amounts than a normal baby. Such infants suffer no nutritional effects Some youngsters may encounter difficulty mainly with liquids while in others the converse will be true, the trouble being with solids The feeding difficulties may lead to aspiration of food particles into the tracheobronchial tree and pneumonitis

esophageal defect at the level of the third or fourth thoracic vertebra (Fig 20B) Tracheography is neither indicated nor helpful in these patients An aortogram will demonstrate the right subclavian artery to arise at a point farther along the aortic arch than is normal (Fig 21)

Surgical Treatment: At thoracotomy and after exposure of the aortic arch and its branches the right subclavian



Fig 20 *A*, Barium swallow examination of a patient with aberrant origin of right subclavian artery *B*, Lateral view showing oblique filling defect high in the thorax on the posterior esophageal wall due to aberrant right subclavian artery

Radiologic Findings: Diagnosis of the malformation depends almost entirely upon performance of contrast visualization of the esophagus for standard chest x-rays show no abnormality The esophagogram usually is pathognomonic of the condition The line of compression runs obliquely upward, from the patient's left toward the right thoracic outlet (Fig 20A) The lateral view demonstrates a small posterior

artery will be seen to arise from the aortic arch distal to the left subclavian artery Temporary occlusion of the vessel while the anesthesiologist palpates the right radial or brachial pulse will aid in positively identifying the correct vessel for ligation Failure to identify the artery properly may lead to catastrophe Once the right subclavian artery is ascertained to be anomalous and to be the cause of the difficulty it



Fig 21: Retouched aortogram performed through the left subclavian artery which demonstrates the aberrant origin of the right subclavian artery

should be doubly clamped and divided preferably after two mattress suture lines have been placed across the vessel by way of the fenestrations in Bailey-Musser clamps (Fig 22A,B)

Results of Surgery: Again the results of operative therapy are extremely satisfying for the mortality approaches zero as does the morbidity The amelioration of symptoms approaches 100 per cent

References

1 Hommel cited by Poynter C W M Arterial anomalies pertaining to aortic arches and branches arising from them Lincoln, Neb., Nebraska Univ Studies 16 220, 1916

2 Bayford, D An Account of a Singular Case of Deglutition. Mem. M Soc London, 2nd Series, 2 275 1794

3 Wolman, I J J Pediat., 14 527 1939

4 Arkin, A., Wien. Arch. inn Med., 12 385 1926.

5 Neuhauser E. D B Am. J Roentgenol 56 1 1946

6. Gross, R. E. New England J Med., 233 586, 1945

7 Bailey C. P Surgery of the Heart, Lea & Febiger Philadelphia, 1955 p 181

8 Gross, R E., The Surgery of Infancy and Childhood, W B Saunders Co., Philadelphia, 1953 p 913

9 Gross, R. E. and Neuhauser E. D B Pediatrics, 7 69 1951

10. Blumenthal S and Ravitch, M M Pediatrics, 20 903 1957

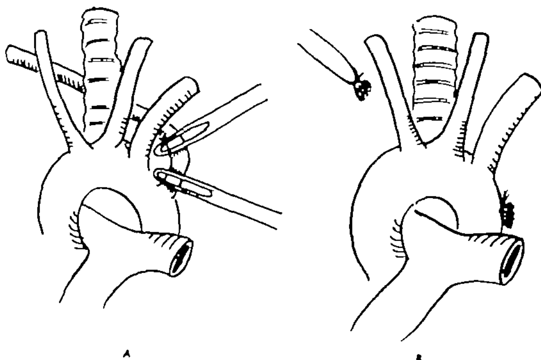


Fig 22: A, Aberrant right subclavian artery at thoracotomy The aberrant artery may be sutured through fenestrated Bailey-Musser clamps and then divided. B, The divided ends of the artery retract widely if properly mobilized during the dissection.

SURGICAL TREATMENT OF VENTRICULAR SEPTAL DEFECTS

CHARLES P BAILEY, M D., AND EDWARD A FITCH, M D

The term ventricular septal defect denotes any direct communication between the right and left ventricles. It may be but a probe-sized perforation in the muscular portion of the septum which closes off during ventricular systole permitting but a minimal shunt. At the other extreme it may amount to a total absence of the septum.

In the majority of instances the ventricular septal defect presents in that portion of the septum which is adjacent to the cusps of the aortic valve (Fig 1) Occasionally there is an associated malformation of the root of the aorta, perhaps with prolapse or low attachment of a valve cusp, with resultant aortic incompetence (Fig 2)

In the condition known as atrioventricularis communis (common atrioventricular canal), there is defective development of both the atrial and the ventricular septum in the region of the atrioventricular valves. One or both of these valves may be malformed and incompetent (Fig 3) In this complicated defect the four cardiac chambers are in direct communication at least during the period of ventricular filling In this variety of malformations the distinction between atrial septal defects of the ostium primum type and atrioventric-

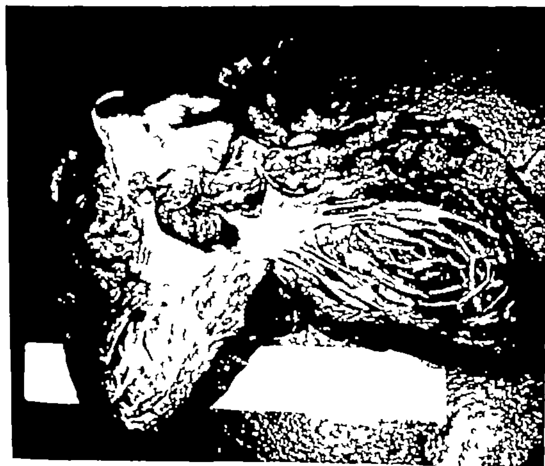


Fig. 1 Large typical ventricular septal defect as seen from the left Note close relationship to aortic valve cusps. A prosthetic patch of Ivalon sponge has bee used for its closure

Fig 2 Malformation of the aortic valve associated with a high ventricular septal defect causing aortic incompetence (Taussig, H B Malformations of the Heart Commonwealth Fund, Cambridge Ed 1, p 401)

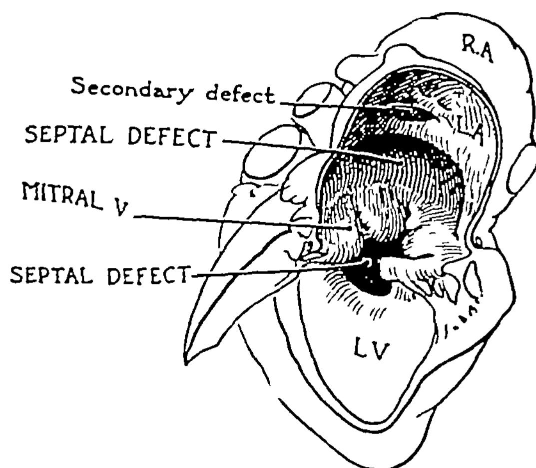


Fig 3 Common atrioventricular canal due to failure of proper union of dorsal and ventral endocardial cushions Both the atrial and the ventricular septae are deficient Usually the atrioventricular valves are malformed (Taussig, H B: Malformations of the Heart Common wealth Fund, Cambridge Ed 1, p 355)

ularis communis often becomes a matter of academic classification

Etiology

Most ventricular septal defects are the result of failure of proper fusion of the upper end of the muscular ventricular septum with the lower end of the aortic septum or with the dorsal and ventral endocardial "cushions" from which are derived the atrioventricular valves Isolated defects of the muscular portion of the septum may be the end-result of a fortuitous union of two large myocardial sinusoids involving opposite sides of the interventricular septum Such communications are con-



Fig 4 Traumatically caused ventricular septal defect in a 17 year old boy who had suffered an auto accident (steering wheel injury) five weeks previously

sidered to be normal in some of the reptilia, notably the python.

Trauma, either of a penetrating or a nonpenetrating type may produce a laceration of the ventricular septum (Fig 4)

The myomalacia of acute myocardial infarction when it involves the septum may eventuate in perforation

Classification

While the various types of ventricular septal defect have been described in many ways [1-4] the classification of Kirklin[5] and associates is admirable in its simplicity and in its applicability to surgical requirements. Table 1 presents it in outline form

TABLE 1

Types of Ventricular Septal Defect Encountered in 36 Surgical Cases

Type of Defect	*Cases*		
Defect related to ventricular outflow tracts		27	
Inferior to crista supraventricularis	25		
Superior to crista supraventricularis	2		"High"
Defect related to ventricular inflow tracts		11	
Beneath septal leaflet of tricuspid valve	8		
Near apex of muscular septum	3		"Low"

(Kirklin, J W, et al J Thoracic Surg, 33 46, 1957)

In this classification the right ventricular outflow tract is considered to extend from the pulmonary valve above to the nearest portion of the tricuspid valve below. The inflow tract of the right ventricle lies posterior and caudal to the outflow tract These two tracts can be considered the straight limbs of a distorted letter "U" which become joined toward the apical region of the right ventricle (Fig 5)



Fig 5 Usual sites of ventricular septal defects as seen from the right 1—High defects above the supraventricular crest, 2—high defects below the supraventricular crest, 3—low defects beneath the septal leaflet of the tricuspid valve, 4—low defects in the apical portion of the muscular septum In repairing defects at locations 2 and 3, there is appreciable risk of injury to the main atrioventricular conduction bundle (His) (Kirklin, J W, et al J Thoracic Surg, 33 46 (Jan) 1957)

Physiologic Effects

In the early uncomplicated case of ventricular septal defect the left intraventricular pressure during systole considerably exceeds that within the right ventricle Therefore, during systole, a portion of the left ventricular blood is expelled into the right ventricle or into the immediately contiguous root of the pulmonary artery It is believed that in this latter situation the violent alternations which are imparted to the pulmonary arterial pressure contribute to a rather rapid degeneration of the pulmonary vascular bed

The magnitude of this left-to-right shunt is directly proportional to the size of the defect and to the pressure differential which exists between the two ventricles, and inversely proportional to the relative resistances of the aortic and pulmonary vascular beds The degree of alignment of each of the great arteries (over-riding) with the ventricular chambers must also be considered Since the end-diastolic pressure within both ventricles normally is zero, no shunting takes place except during systole Hence, the magnitude of the shunt through a defect of the ventricular septum tends to be smaller than

spective tendencies toward a right-to-left and toward a left-to-right shunt countering each other

A still further advance in the pulmonary arterial deterioration will result in such an increase in the total pulmonary arterial resistance that an overt right-to-left shunt will occur through the defect This produces undersaturation of the systemic arterial blood and usually is manifested by visible generalized cyanosis

It must be pointed out that in the case of a very large defect of the ventricular septum the respective right and left ventricular and arterial pressures necessarily must have been equalized from birth Without a *physiologic* elevation of the pulmonary vascular resistance early in life nearly all of the systemic blood would have been shunted into the lungs with consequent failure of the general bodily circulation and death This has been attributed by Eldridge, *et al*,[8] to a persistence of the normal *fetal vasculature* of the lung With large septal defects a certain amount of mixing of the blood between the respective sides of the heart necessarily must take place and a certain degree of undersaturation of the arterial blood in these cases may be expected However, this does not have the same untoward significance as does a more recent appearance of cyanosis in an originally noncyanotic patient with a smaller ventricular septal defect in whom occlusive vascular pulmonary disease presumably has developed Histologic examination of the lung tissue from infants with very large defects, therefore, may not reveal the usual form of vascular degeneration but, rather, a persistence of the normal *fetal vasculature*

Symptoms

Symptomatology in ventricular septal defects varies greatly and is proportional to the size of the defect, to the magnitude and direction of the shunt, and to the level of the pulmonary hypertension, if any

Very small defects usually are seen in the completely asymptomatic individual who can perform strenuous physical activity and lives out a full life's span (Roger's disease) The only evidence of abnormality may be a loud systolic murmur and perhaps a minimal amount of exercise intolerance Palpitation is common

Larger defects may be asymptomatic until the secondary effects of pulmonary and right ventricular hypertension such as cyanosis (due to right-to-left shunting), right heart strain (and frequently left) and failure develop Easy fatigability, retardation of bodily growth, shortness of breath, and later cyanosis and clubbing of the fingernails and toenails may be noted and tend to progress

Patients with very large defects (approaching the status of a single ventricle) may be severely symptomatic from birth. A degree of over-riding of the aortic root may be present and exaggerate the tendency toward right-to-left shunting

None of these symptoms or signs is pathognomonic for the definitive diagnosis of interventricular septal defect. Symptoms may be absent from birth, may be present from birth, or may appear at any age This type of cardiac defect, however, usually causes death The final episode may be a bout of subacute bacterial endocarditis, intercurrent pulmonary infection, Adams-Stokes syndrome (heart block), right heart failure, or sudden (hypoxic) death

Physical Signs

Infants with ventricular septal defects usually present a rapid vigorous cardiac action and a loud harsh systolic murmur distributed generally over the precordium and usually maximal over the lower portion of the sternum Unlike the rather similar murmur of pulmonic stenosis it is accompanied by an accentuated pulmonary second sound. Not infrequently a palpable thrill accompanies the murmur A diastolic basal murmur should alert one to the possibility of concomitant aortic insufficiency which may reflect an abnormality of cusp development.

In approximately 20 per cent of these individuals a deformity of the thorax in the region of the precordium will be apparent, the left side of the chest protruding This is thought to be caused by overdevelopment of the right ventricle

Cyanosis or arterial undersaturation is unusual except in the three following situations It occurs in persons with advanced pulmonary vascular changes as a late phenomenon, in those with over riding of the septum by the aorta, and in those with very large defects which permit an element of physical mixing of the blood from the two ventricles In these latter two situations a portion of the unsaturated blood from the right ventricle will enter the aorta to increase the amount of reduced hemoglobin within the arterial blood. In such cases cyanosis will have existed from birth.

Not infrequently in patients with ventricular septal defects the lung fields, especially the bases posteriorly will present rales or rhonchi. These auscultatory phenomena usually are related to the presence of an acute or subacute respiratory infection, to which these patients are especially prone, rather than to the increased vascular congestion, *per se*

Diagnostic Procedures

Radiographic Examination: The standard 72 inch posteroanterior roentgenogram usually will reveal a degree of overall cardiac enlargement varying from slight to great (Fig 7) The

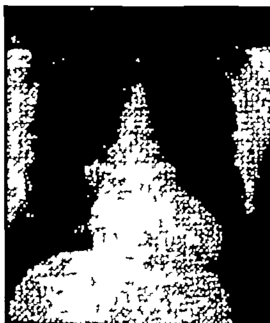


Fig 7 Posteroanterior roentgenogram. A 29 year old white male with ventricular septal defect Note the increased pulmonary vascular markings. The *pulmonary conus* seems less prominent than is usual in these cases.

region of the pulmonary conus will be prominent. The pulmonary vascular markings may be accentuated perhaps to an extreme degree.

Angiocardiography Venous angiocardiography will reveal no abnormal circulatory pathway in a patient with a left to-right shunt through a septal defect but may emphasize the dilatation and congestion of the pulmonary vascular bed. When a right to-left shunt exists, the opaque material will outline the course of the blood as it passes from the right ventricle through the defect and into the aorta (Fig 8A B)

Ventriculography (Left): Direct needle puncture (by the subxyphoid route) of the left ventricular cavity will permit opacification of this chamber and may show dye escaping into the right ventricle in a patient with a left-to-right shunt (Fig 9A,B,C)

Dye Dilution Studies: The use of various dyes which can be detected by colorimetric methods provides a valuable adjunct to the diagnosis of intracardiac foramina[9, 10] and the quantitation of the concomitant shunts. Several methods are available, all involve the in appropriately obtained blood samples.[11]

Specifically, in the case of an interventricular septal defect with a left-to-right shunt, dye is injected into a lobar pulmonary artery through an inlying cardiac catheter. In the presence of such a defect samples drawn from the right ventricular chamber will show a sudden abnormal rise in dye concentration. Samples taken at points proximal to the defect will show no dye until a complete circulatory circuit has been made

In a similar manner, the right-to-left

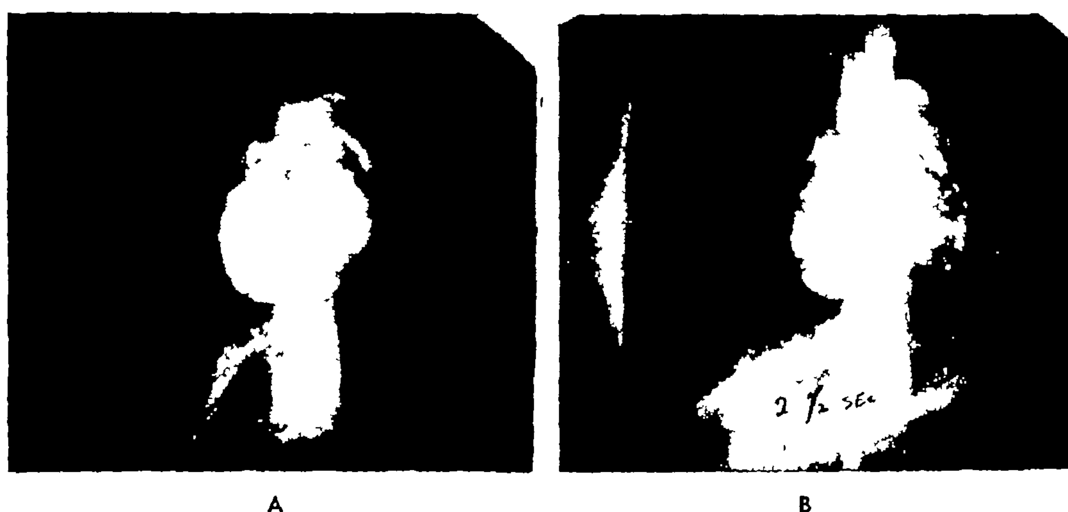


Fig 8 Angiographic visualization of the flow pattern in an 18 year old white female with a ventricular septal defect and a right-to-left shunt Note simultaneous opacification of the aorta and the ramifications of the pulmonary artery

rapid injection of a quantity of dye into the cardiovascular system, and the recording of its transit at another point "downstream" in the vascular conduit Monitoring of the dye in transit at the point of determination can be accomplished by a constant, steady withdrawal of blood through a photoelectric cell, or by the direct photoelectric registration of the dyed blood as it passes through a superficial "transparent" tissue (such as the earlobe) Similar methods have been utilized which employ radioactive serum albumin and monitor the radioactivity component of a septal shunt can be demonstrated by injection of the dye at a point proximal to the defect (peripheral vein or right atrium) and notation of its unusually prompt detection within a systemic artery

By correlation of the dye dosage and the characteristics of the concentration curve, the amount of blood involved in the shunt (regardless of direction) can be calculated The cardiac output readily may be obtained simultaneously

Catheterization of the Right Heart: A Cournand or other suitable catheter introduced by way of a systemic vein

toward the heart will enter the right atrium, pass through the tricuspid valve traverse the right ventricular chamber and enter the pulmonary artery In the presence of a ventricular septal defect the tip actually may traverse the defect to enter the left ventricle or the aorta (Fig 10)

Blood samples obtained at various points during advancement of the catheter tip will reveal a "step-up" in the oxygen saturation at the ventricular level (Fig 11A) unless complete reversal of the shunt has taken place. If the direction of the shunt has become reversed or bidirectional, a reduction



Fig 9 Demonstration of a left-to-right shunt through a ventricular septal defect by direct injection of contrast substance into the left ventricle A, Left ventricular chamber filling with dye by a pressure injector technic. B Beginning filling of right ventricle and demonstration of communication through a defect in the region of the outflow tract C Right ventricle opacified

in the oxygen saturation of the femoral (or brachial) arterial blood samples will be noted. However there usually will be some "step-up" in the oxygen saturation at the ventricular level (Fig 11B)

The pressure levels in the various portions of the vascular pathway often will reveal an elevation in the right ventricular and pulmonary arterial tension if the defect is large or if significant pulmonary arterial change has occurred (Fig 11A,B)



Fig. 10: Passage of the cardiac catheter from the right ventricle into the aorta. When truncus arteriosus and transposition can be ruled out this establishes the presence of a ventricular septal defect

Clinical Course

The outlook for infants born with defects of the ventricular septum is extremely variable. It has been estimated that more than 50 per cent of these patients do not survive the first year[12] Failure to gain weight and to develop properly an extremely rapid heart rate, rapid respiration, and a pro-

nounced predisposition to respiratory infections characterize the course in many such infants

Other individuals apparently develop normally and are able to compete physically with their fellows for many years Some remain well and active for a normal life span (Roger's Disease)

The classical patient follows an intermediate course While troubled by palpitation and a somewhat limited exercise tolerance, he survives his respiratory difficulties and the period of infancy, and enjoys a relatively normal period of childhood or young adult life until degeneration of the pulmonary arterioles becomes pronounced Thereafter, shortness of breath and cyanosis upon extreme physical activity gradually become replaced by shortness of breath with less and less activity and, finally, by cyanosis even duri

While the average span of certain, obviously it is cor shorter than normal In the onset of subacute bacterial often has contributed to episode. Today, with the avail effective antibiotic therapy de: this cause are rare Most deat may be attributed to circulator either congestive or asphyxial

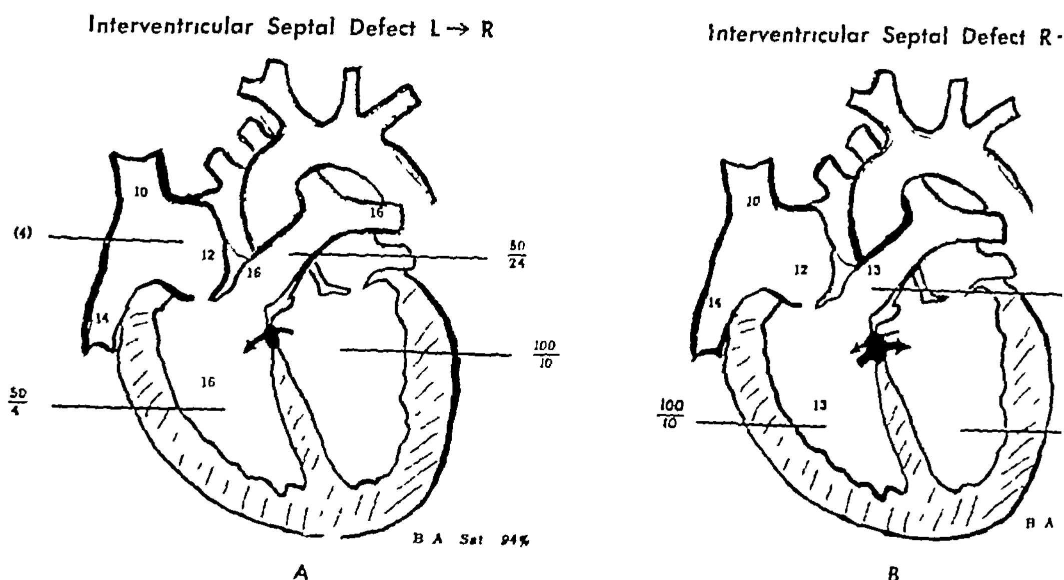


Fig 11 Typical findings during cardiac catheterization (right) in ventricular septal The numbers within the chambers indicate the oxygen content of the blood in volumes pe The outside numbers depict pressure readings (mm) within the respective chambers A, presence of a left-to-right shunt a "step up" will be noted in the oxygen content of blood the right ventricle over that within the right atrium The right ventricular and pulmonary a pressures may be normal, may be elevated to the level of those within the left ventricl aorta (pulmonary hypertension), or may be measured at an intermediate range The saturat the peripheral arterial blood will be within normal limits B, In a case with pulmonary tension and a predominant right-to left shunt there usually will be an element of biphasic shu Therefore, there will be some "step up" in the oxygen content of the right ventricular blood though the brachial arterial samples show undersaturation The right ventricular and pulm arterial pressures will be at systemic levels Patients in this condition usually are cons inoperable

Undoubtedly, the fate of th tients is dependent upon various priate interrelations between t and location of the defect, the ence or absence of a residuum normal "fetal" vascular resistan presence or lack of "over-ridi the aorta, and perhaps certain tously associated lesions Undou such intangible individual factor

vitality of the myocardium and the ability of the pulmonary arterioles to withstand prolonged active congestion must play a significant part in the final outcome.

Surgical Therapy

Due to the complex anatomical and functional relations with contiguous structure no truly effective means for the closure of ventricular septal defects could be worked out before the advent of direct vision surgery. Several authorities have reported technics of repair using closed technics but none can be considered to have been truly successful.[11-15]

General bodily hypothermia allowed an attack upon the defect under direct vision but the permissible operating time was severely restricted and the surgical mortality was high.[16-19]

The gradual perfection of the various extracorporeal circulatory systems[20-25] and the use of plastic prosthetics[26] (*nylon, teflon, dacron, marlex*) for the repair of larger defects have rendered open correction of these lesions progressively safer. However world experience with these operations to date has been attended by a relatively high mortality.[27] The reasons for this mortality seem clear enough. Some surgeons continue to operate upon end stage patients in whom the pulmonary vascular bed has been destroyed irreparably. Others attempt fully corrective operations upon very tiny infants in poor condition in whom the operative risk is extremely high.[27] Many surgeons continue to overlook the possibility of precipitating severe acute alterations in the intracardiac pressure relationships by closing a defect which may be essential (at least temporarily) for the maintenance of a long established although admittedly abnormal hydrodynamic situation. Heart block due to suture strangulation of the atrio-ventricular conduction system (Bundle of His) continues to contribute an increment of mortality and morbidity.

Lung Biopsy: While one can hardly fail to be sympathetic to the surgeon who attempts to salvage a borderline patient with severe pulmonary hypertension it is becoming increasingly apparent that some reasonable *line of demarcation* must be established. Preoperative pulmonary biopsy although contested by some, would seem to offer the most reliable means of establishing the presence or absence of irreversible pathology of the pulmonary vascular bed.[28]

Surgery in Infancy: In the case of very ill infants who do not tolerate total circulatory bypass well there exists a highly effective and reasonably safe temporizing method. "Banding" of the pulmonary artery to reduce its caliber has been practiced by Muller and Damman,[29] Therkelsen, *et al*,[30] and many others. The surgical narrowing of the lumen of the main pulmonary artery by external constriction significantly reduces the pressure which prevails within the distal pulmonary arterial system, increases the pressure in the proximal portion of the pulmonary artery and right ventricle and reduces greatly or even abolishes the left-to-right shunt through the defect. In other words it overcomes nearly all of the untoward physiologic effects of a ventricular septal defect although the right ventricle necessarily continues to remain in dynamic competition with the left. This, however it is capable of doing for many years before congestive heart failure develops.

By so reducing the shunt these infants can be "carried along" to an age and size at which open definitive sur

gery can be carried out with every prospect of success. Probably corrective operation should not be delayed much past the second birthday. Sirak, Hosier, and Clatworthy[31] have reported a case which developed progressive vascular narrowing at the site of "banding," resulting eventually in a right-to-left shunt through the defect thus establishing a hydrodynamic pattern identical to that seen in the tetralogy of Fallot.

Prevention of Severe Intracardiac Pressure Alterations: Kolff and Effler[32] were the first to report the prompt development of high pressures within the left atrium following the complete closure of ventricular septal defects. In many instances this was attended by the development of acute hemorrhagic pulmonary edema and death They have proposed direct catheter drainage of the left atrium to avert this complication Undoubtedly, there are less severe stages of this phenomenon which may be manifested during the postoperative period by pulmonary congestion, excessive secretion of bronchial mucus, sudden cardiac arrhythmia, or heart failure Experience with certain of these complications has led to speculation as to possible methods by which the pressures within the various cardiovascular "compartments" might be maintained within safe limits Sirak and Hosier[33] have suggested that an artificial patent ductus arteriosus might be created at the time of closure of the septal defect. Lillehei has made the suggestion,[34] based upon Lewis'[35] experimental demonstration of the propensity for small "holes" in a patch of ivalon sponge used to repair an atrial septal defect to close off spontaneously, that ventricular septal defects might be treated more safely with a multiply perforated patch He particularly suggested the use of such a prosthesis in patients with severe pulmonary hypertension. However, it has been found that small perforations in such a patch become closed off within a few hours[36] Thus little more protection is afforded by this technic than is obtained by Kollf's prolonged catheter drainage of

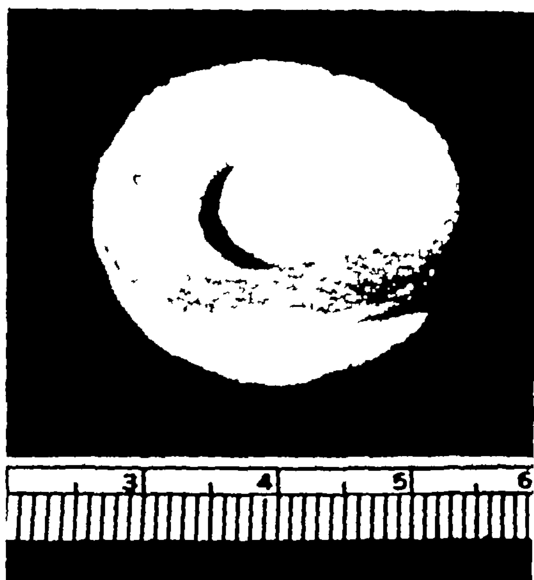


Fig 12 "Prosthetic foraminal valve" prepared by subtotally slicing a slab of compressed (and teflon reinforced) ivalon sponge, and then creating a central opening in one of the "leaves" (Larios, R, Fitch, E A, Blanco, G, and Bailey, C P)

the left atrium Large perforations in such a prosthetic may not close off at all Cooley[37] has reported two cases in which ivalon patches used to repair ventricular septal defects became ruptured within a year. It would seem, therefore, in order to insure permanent closure of ventricular septal defects that a very strong imperforate prosthetic patch is essential

Larios, Fitch, Blanco, and Bailey[38] have reported their early experiences with the use of a prosthetic valvular patch (Fig 12) of *teflon reinforced ivalon* for the closure of septal defects. Such prosthetic "foraminal valves" which are fashioned in the form of a miniature "toilet seat," may be placed

in such a manner as to permit early continuation of a shunt in a unidirectional fashion (Fig 13A,B) The "lid" of the prosthesis gradually becomes adherent to the *ring* thus closing the defect completely within eight to twelve weeks (Fig 14) It would seem that application of this principle should afford sufficient time for gradual adjustment left and right ventricular pressures might tend to keep the prosthetic valve open and to prevent approximation of the hinged flap to the ring if so placed as to permit continuation of a left-to-right shunt (Fig 13B) It is our present feeling that such prosthetic "foraminal valves" should be applied only in such a manner as to permit the

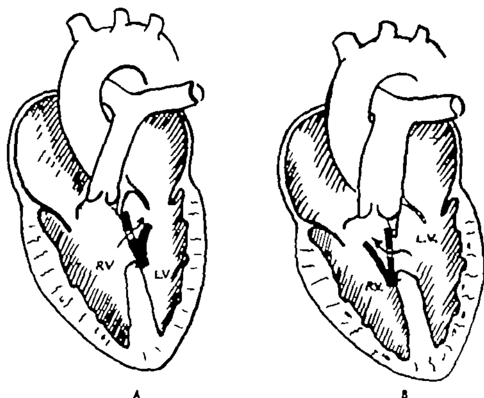


Fig 13 Schematic illustration of the methods of placement of the prosthetic foraminal valve to permit temporary unidirectional shunting A, Placement which permits right-to-left shunting The relative disparity in the respective intraventricular pressure levels favors closure of the valve and eventual obliteration of the passage B Placement which permits left-to-right shunting This method of application is frowned upon since the normal disparity in the respective ventricular pressures tends to keep the valve open and, hence to perpetuate the shunt

of the heart to the new pattern of the circulation.

Theoretical and practical disadvantages have caused us to continue our search for other methods to insure maintenance of proper postoperative pressure relationships. It is difficult to fashion a functional prosthetic valve for certain small or irregularly shaped defects. One must accept the likelihood that the natural disparity between the occurrence of a right to-left shunt whenever desirable or necessary (Fig 13A) Hence, they seem suitable for the gradual closure of ventricular septal defects in patients with the tetralogy of Fallot and, perhaps, in pulmonary hypertension.

Our experience with the successful prevention of acute pulmonary edema during and following surgery for mitral stenosis[39, 40] by the elective creation of

an atrial septal defect of small (7 mm) diameter caused us to try a similar procedure in certain patients with ventricular septal defects We know from postoperative catheterization of our patients with surgically corrected mitral stenosis that these created atrial septal defects consistently close off within a short period of time (from three to twelve weeks). It was felt that applica-

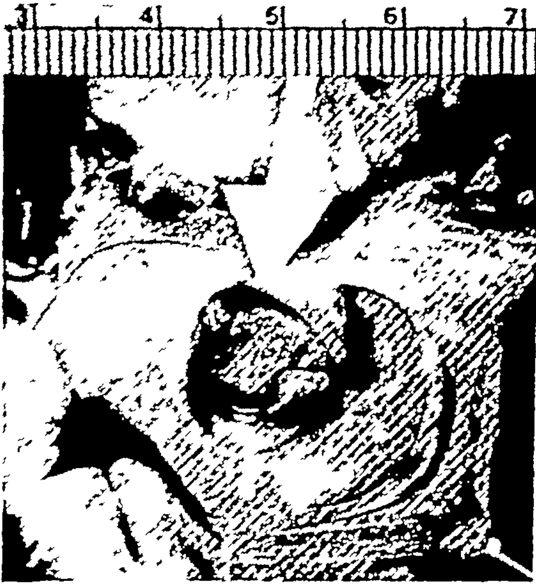


Fig 14 Animal specimen, typical findings with respect to the prosthetic foraminal valve several weeks after implantation in the favored manner (Fig 13A) The two leaves of the prosthetic valve have become agglutinated by enveloping and infiltrating fibrin Ultimate organization will convert the structure into a fibrous scar with a plastic "skeleton"

tion of this technic in cases with ventricular septal defects likewise would afford sufficient time for the heart to become adequately adjusted to the altered circulatory situation. The operative creation of such a defect, furthermore, is simple and safe. It provides automatic "balancing" of the respective intra-atrial pressures regardless of the direction of the preoperative shunt (right-to-left, left-to-right, equalized, or bi-directional). Today, except for the closure of very small ventricular septal defects we use this method routinely, and are most pleased with the improvement in the initial response. We have not had a single operative death or significant postoperative morbidity which could in any way be attributed to the creation or presence of such a created atrial septal defect.

While the subsequently described operative technics are our own they do not differ in principle or concept from those used by others

Surgical Technics. ***Banding of the Pulmonary Artery.*** When an infant is deemed too

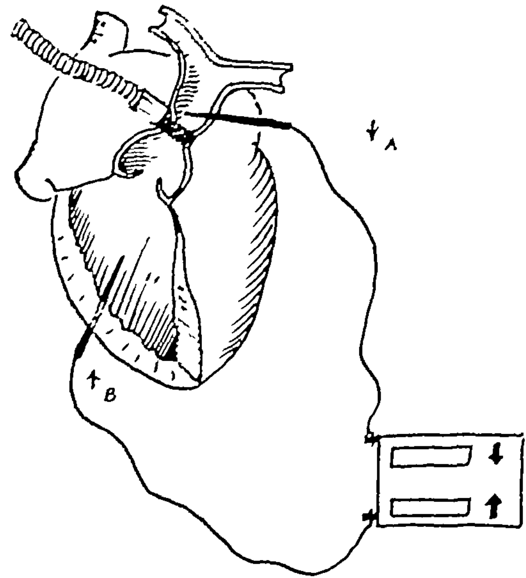


Fig 15 "Banding" or constricting the pulmonary artery to reduce the magnitude of a large left-to right shunt Simultaneous pressure recordings are determined by direct needle puncture of the right ventricle and the pulmonary artery

delicate or fragile for a major corrective intervention, the pulmonary artery may be narrowed or "banded" to increase the total pulmonary vascular resistance, thus reducing the amount of the left-to-right shunt

An anterolateral incision is made in the third left intercostal space The pericardium is incised longitudinally anterior to and parallel with the left phrenic nerve. The pericardial incision may be enlarged if necessary by a medial extension to expose the first portion of the pulmonary artery

The aorta and pulmonary artery are separated by blunt dissection at a level just above the conus ligament An umbilical tape is passed about the pulmonary artery and is

incorporated within a Rumel Belmont tourniquet.

Two needles connected with a multichannel pressure recorder are inserted respectively into the right ventricle and the distal pulmonary artery (Fig 15) Pressure tracings are obtained. The tourniquet is tightened progressively narrowing the pulmonary artery. When the distal pulmonary arterial pressure has been lowered to normal or to as low a hypertensive level as is deemed safe, the tape is tied in such a manner as to maintain this degree of constriction.

Open Heart Surgery Circulatory Bypass Many types of pump-oxygenator systems have been designed and used for the correction of ventricular septal defects.[a] While saw However the vibrating Stryker saw (Fig 16) which readily and smoothly divides hard structures such as bone while sparing soft structures with which it may come in contact provides a much safer and seemingly nearly ideal method for separating the sternum longitudinally In a small child with a cartilaginous sternum scalpel and scissors alone may suffice. Bone wax applied to the cancellous sternal margins will restrain bleeding from the bone marrow Application of the electrocoagulating current to the upper and lower periosteal cut edges will prevent persistent oozing from these sites after the establishment of incoagulability of the blood.

If before surgical division of the sternum, the under surface is separated digitally from

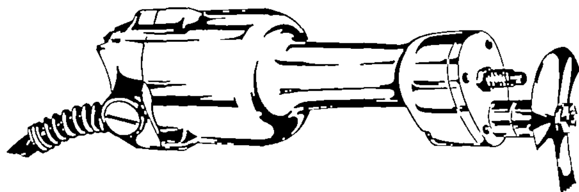


Fig 16: Stryker Vibrating Saw

individual advantages and disadvantages have been attributed to each, it is fair to say that nearly any of those which currently are available, if properly used are capable of providing safe total cardiopulmonary bypass for periods up to one hour in duration. Elective cooling of the patient with a heat exchanger as advocated by Brown and Sealey[a] may permit a longer period of safe bypass. However most cardiac surgeons do not combine hypothermia with bypass as a routine procedure

Opening the Chest While many cardiac surgeons continue to transect the sternum and to open both pleural cavities for maximal exposure of the heart, we along with many of the most experienced have come to believe that the sternal splitting incision provides adequate surgical exposure while preserving maximal functional respiratory reserve This latter thoracic exposure is performed through a midline longitudinal incision. The sternum may be divided by a Lebsche knife, Shumacker sternal shears, or a rotating circular the mediastinal tissues there will be very little risk of entering either pleural cavity during this phase of the procedure.

Opening the Pericardium. Once the sternal halves have been separated (preferably with Morse sternal spreaders) (Fig 17) the operator may visualize the pericardial surface and

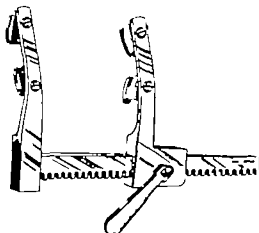


Fig 17 Morse d ble retractor for separation of edges of longitudinally divided st rnum.

can gently separate the pleural reflections and the respective lobes of the thymus gland from it The pericardium then may be incised from the most cephalic extension at its aortic reflection to the level of the diaphragm The cut edges may be coagulated with the high frequency current If these edges are sutured immediately to the respective sternal halves, the likelihood of inadvertent laceration of either pleural membrane will be reduced

Cannulation of the Cavae The patient's blood is rendered incoagulable by the intravenous administration of heparin sulfate in a dosage of 3 0 mg per kilogram of body weight Ten minutes are permitted to elapse for attainment of the desired anticoagulant effect Meanwhile the superior and inferior venae cavae are dissected free of pericardial attachment by sharp and blunt separation with a right angled clamp They are encircled with umbilical tapes The ends of the latter are passed through moderately stiff segments of rubber tubing which are cut long enough to protrude from the operative incision

The right auricular appendage is excluded from the circulation by a noncrushing clamp Two separate areas upon the appendageal surface are selected for the application of purse string sutures of fine strong material Triple zero nylon sutures swaged within curved round needles are quite satisfactory for this purpose An incision just equal to the diameter of the particular multiwindowed plastic catheter chosen for cannulation of the superior vena cava is made within one of these suture circumscribed sites The lips of this incision are picked up and the tip of the selected catheter with the appropriate sized Fitch[13] semi-malleable obturator in place is inserted snugly into the tiny incision (Fig 18) As the excluding clamp is removed from the appendage the catheter is so directed and advanced that its tip (and all of its terminal perforations) comes to lie within the lumen of the superior vena cava, well above the level of the encircling umbilical tape The obturator is removed from the catheter, venous blood samples are taken for chemical and cytologic studies and a clamp is applied across the catheter terminally The purse string suture is tied securely, passed twice around the catheter, and tied again This latter maneuver, then, is repeated to guarantee secure fixation to the atrial wall

The portion of the appendage which contains the remaining purse string is excluded with a smaller clamp A similar incision is made within the encircling suture line and the inferior vena cava is cannulated with a somewhat larger multiwindowed plastic catheter Fixation is similar



Fig 18 Fitch obturator (one of a graduated series) below multifenestrated Bardic catheter Obturator within Bardic catheter

Arterial Cannulation While some cardiac surgeons continue to utilize the left subclavian artery for the perfusion of the arterial tree, most of them now use one of the considerably larger femoral arteries After considerable experience the authors prefer to use both femoral arteries since the double passageway reduces the pressure gradient across the arterial cannulae (and consequently the blood trauma) significantly These cannulations are performed simultaneously by a separate surgical team using bilateral inguinal incisions Transverse partial division of these sizeable arteries facilitates nonconstrictive vascular repair upon termination of the perfusion

In small children, because of the diminutive

size of the peripheral arteries, it has become our custom to insert an arterial catheter directly into the lumen of the ascending aorta. A purse string suture of fine strong material (2-0 nylon) is placed deeply within the adventitia at a selected site upon the anterior wall of the ascending aorta. This area is excluded from the remainder of the aortic lumen with a suitable clamp (Beck or Potts type of dentate clamp) An incision equal to the diameter of the selected plastic catheter is made into the aortic wall within the confines of the purse string suture. Its lips are picked up and the bevelled end of a somewhat shortened catheter is inserted into the excluded portion of the arterial lumen. At a suitable signal the clamp is removed, the catheter is advanced into the aortic arch, and the purse string suture is tightened and tied (Fig 19A,B,C,D) The suture ends are used to doubly encircle the catheter at two separate levels before tying

Monitoring Proper monitoring of the peripheral arterial and venous pressures is fundamental to the survival of the patient. By insertion of fine polyethylene or other type of tubing into an artery and a vein, both the mean and actual pressure variations may be determined continuously throughout the period of perfusion and for a time after its termination. We believe that at normothermic temperatures the mean arterial pressure level should not be permitted to fall below 65 mm. of mercury nor to rise above 100 mm. The venous pressure should not be below 5 mm. Hg or above 25 These pressure levels are

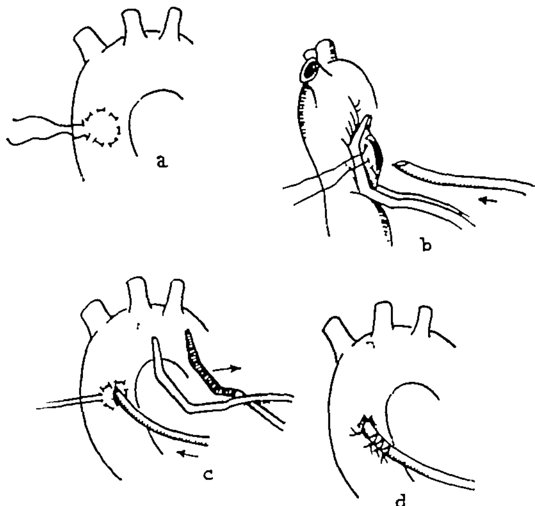


Fig 19: Steps in cannulation of the ascending aorta for arterial perfusion: *a* A purse string suture is placed within the outer layers of the aortic wall. *b*, A portion of the aortic wall (purse stringed) is excluded with a dentate clamp. An incision just sufficient to admit the selected arterial catheter is made within the confines of the suture *c*, The bevelled tip of the catheter is inserted between the lips of the incision and is advanced into the arch as the clamp is removed *d* Tightening of the purse string provides complete hemostasis. The ends are tied about the catheter several times to secure it.

most effectively regulated by adjusting the volume of blood within the patient's circulatory system However, an increase in the rate of pumping, or the administration of vasopressor agents may be used to help elevate the arterial blood pressure

Needless to say, continuous electrocardiographic monitoring is basic to proper control of the cardiac rhythm.[44] Not infrequently electroencephalographic recordings may be useful[45] in detecting the earliest phases of cerebral anoxia While continuously recorded or frequent oxygen saturation determinations

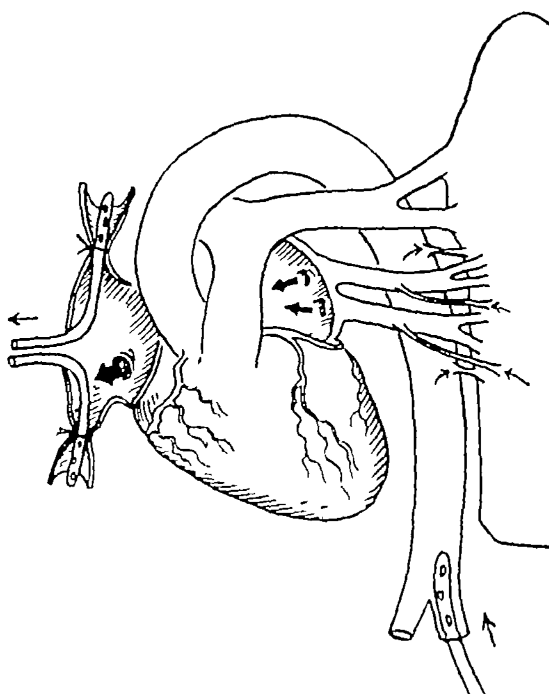


Fig 20 Illustration of chief routes of blood entrance into the heart after cannulation and tape constriction of the venae cavae Coronary sinus flow and bronchial arterial flow continue during circulatory bypass

and *p*H studies may be helpful during the course of the perfusion, with a properly functioning mechanical unit they do not appear to be essential

Compatible Blood for Bypass. While compatible freshly drawn heparinized blood is ideal for circulatory bypass, sometimes this is difficult to obtain especially for patients with the rarer types of blood grouping. The next best choice is heparinized blood not over thirty hours old The use of *edglucate* preserved blood in plastic bags which may be stored for as long as five days has been advocated by Smith, Brown, and Eadie,[46] and has proved satisfactory in our hands

While formerly the perfusion blood was warmed to 37° C (98 6° F.) before using, it now seems preferable to take it directly from the blood bank icebox, thereby simultaneously establishing a mild degree of hypothermia

Establishment of the Perfusion· The arterial cannula must be connected with the arterial tubing from the pump with the greatest of care to exclude even the most minute bubble of air If a suitable bubble trap or a bubble type oxygenator is incorporated within the perfusion circuit less care may be taken with the venous catheters All tubing connections are wired or coupled securely with a suitable mechanical device

Once the connections with the extracorporeal system have been completed it is our practice to remove the obstructing clamps and to establish the perfusion at a low rate of flow Gradually this is increased to take over the entire circulatory load Then the tourniquets (tapes within the rubber cuffs) about the venae cavae are tightened rendering the cardiopulmonary bypass complete (except for the coronary and the bronchial arterial circulations) (Fig 20)

It must be pointed out that many cardiac surgeons prefer to start the perfusion initially at a predetermined (estimated full) rate and to tighten the tourniquets at once This latter technic is especially useful in patients with aortic incompetence, in whom the danger of inadvertent distention of the left ventricle by continuous flow perfusion during the period of diastole is ever present during such bypass

Incision of the Ventricle While transatrial,[3] left ventricular[47] and pari-aortic[48] approaches to defects of the ventricular septum have been proposed and tried, it now is accepted generally that, in the majority of cases, only a large opening into the right ventricular chamber will disclose the pathology completely and at the same time permit its efficient correction A full thickness incision of the right ventricular wall, which extends from the pulmonary valve above to within a centimeter of the ventricular apex, will provide maximal facility both for orientation and instrumental manipulation Due care should be taken to keep the line of incision at a safe distance from the anterior descending branch of the left coronary artery lest it be encircled by the reparative sutures Large branches of the right coronary artery should be preserved, if possible, by appropriate deviation of the line of the incision or by

limiting its extent (if this is consistent with the surgical requirements)

Usually by the use of an appropriate low pressure suction system the right ventricular chamber can be kept emptied of blood so that visualization of the septal anatomy is adequate If the coronary venous return proves to be excessive, the root of the aorta may be cross-clamped temporarily abolishing all coronary blood flow It is our practice unhappy experiences encountered in attempting to restore effective cardiac contractions after its employment, have caused many to abandon elective cardiac arrest for the repair of ventricular septal defects.

The bronchial circulation rarely is developed sufficiently to cause excessive bleeding in patients with uncomplicated ventricular septal defects.

Atrioventricular Conduction System: In low

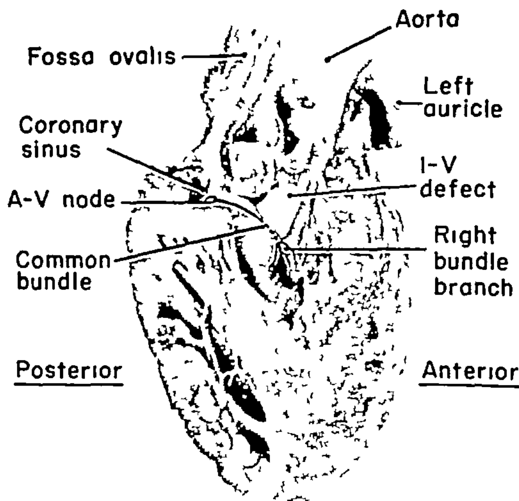


Fig 21 Course of the common atrioventricular conduction system in relation to the dorsocaudal aspect of the usual ventricular septal defect (Truex, R. C., et al.: J Thoracic Surg 35 421 (Apr.) 1958.)

when operating at normothermic conditions to remove the clamp for at least five minutes after a ten minute period of coronary interruption in order to preserve the integrity and tone of the myocardium Thus, by intermittent aortic cross-clamping a sufficient total operating period can be obtained.

Elective cardiac arrest with potassium salts or acetylcholine would seem to avoid this difficulty with coronary venous return. However recent reports as to toxicity* and certain (muscular) defects and in defects occurring above the supraventricular crest no important portions of the conduction system lie in close contiguity to their margins. Therefore, no unusual care need be taken in this respect during the reparative maneuvers.

In the very common outflow tract defects which lie below the level of the supraventricular crest, Truex* has delineated the usual course of the main conduction tract (Bundle of His) and its branches (Fig 21) It readily

may be perceived that the risk of encircling or piercing the main conduction bundle is appreciable only when suturing in the region of the dorsal-caudal quadrant of the defect It is our feeling that tangential passage of the suturing needle on the left ventricular aspect of the defect in this area offers maximal protection against the possibility of causing complete injury of even a superficially placed employed in this region (Fig 22A,B) Indeed there would seem to be no theoretical objection to using such a stitch for suturing the remainder of the margin of the defect

The relationship of the conduction system to the margins of inflow tract defects which lie behind the septal leaflet of the tricuspid valve has not been established so clearly. Not infrequently the tricuspid annulus fibrosus

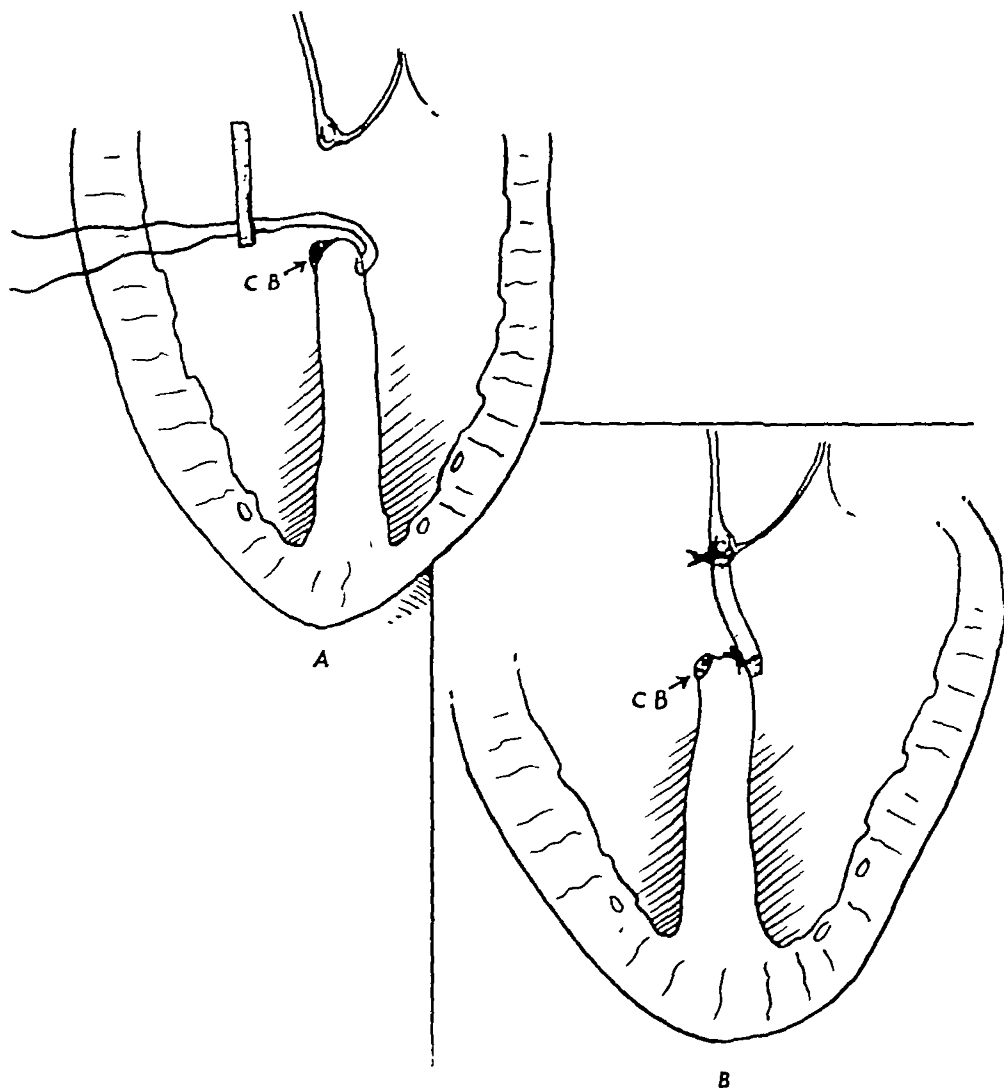
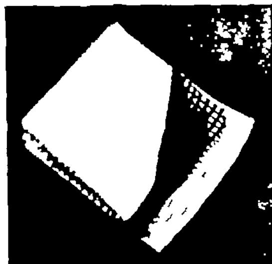


Fig 22 Method of suturing upon the left ventricular side of the margin of the septal defect which avoids the common conduction Bundle C B (here shown in cross section) A, Passage of suture before placement of prosthetic patch B, Patch placed

Bundle of His Even physical entrance of the tangentially passed needle into the substance of the bundle would not necessarily be disastrous (although edema due to the trauma could cause temporary heart block) For this reason we have established the dictum at our clinic that only a mattress type of suture which picks up the left ventricular aspect of the septum in this tangential fashion may be makes up a portion of the dorsal margin of such a defect, and this suggests that the main conduction bundle must lie in close proximity Again it is felt that only a tangential type of suture passed through the left ventricular aspect of the marginal tissue in the dorso-caudal quadrant of such a defect can guarantee against the inadvertent causation of complete atrioventricular heart block

Closure of the Defect: While very small defects can be closed satisfactorily by a direct suture technic, in larger defects it may be preferred to use a prosthetic plastic patch since the suture line then can be established without tension. Most surgeons prefer to use a patch of moderately compressed formalinized polyvinyl plastic sponge (*ivalon*) for this purpose. Others prefer a plastic of greater strength and longer life such as a loosely woven mesh of *teflon, dacron, marlex,* etc. At our own clinic we use either the described prosthetic *foraminal valve* or a



C

Fig 22: C Sandwich-like structure of prosthetic patch, a teflon mesh compressed between two slabs of ivalon sponge.

sandwich prepared from two one centimeter thick slabs of ivalon sponge compressed six fold upon an intervening mesh of teflon (Fig 22C) Thus one obtains in the prosthesis the ease of handling and suturing which ivalon provides along with the integral strength and longevity of teflon.

It is our practice to "tailor" the patch to a size which is slightly smaller than that of the defect, and to a shape which approximates it. The most *dangerous* and difficult portion of the repair is carried out first. In the outflow tract defects which lie below the supraventricular crest, and in defects lying beneath the septal leaflet of the tricuspid valve the portion of the margin near which the conduction bundle may pass is repaired first using the previously described "tangential suturing technic taking care to apply the sutures to the left ventricular side of the marginal tissue. After this line of sutures has been extended beyond the dangerous zone, either interrupted simple or mattress sutures may be used to complete the repair It may be necessary to divide the ventricular attachment of the chordal or papillary support of the septal leaflet of the tricuspid valve in order to gain access to the deepest portion of the defect. The integrity of the tricuspid valve mechanism then must be restored after closure of the defect.

Just before tying down the last sutures in the closure of the septal defect an attempt is made to express any entrapped air from the left ventricle. This may be accomplished readily by permitting the left ventricle to become filled with blood or saline. The last suture is then tightened at the height of ventricular systole If any residual leak persists one or two additional stitches may be placed to complete the closure. If the ventricles are not contracting, being in a state of arrest or ventricular fibrillation, after the defect is closed securely the apex of the

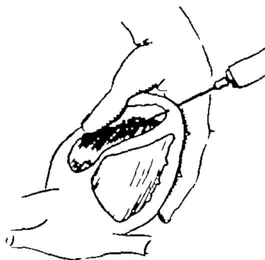


Fig 23 By elevating the apex of the left ventricle any entrapped air may be aspirated in its entirety

heart is elevated and a #17 needle is inserted into the left ventricular chamber contradependently in order to aspirate any entrapped air (Fig 23)

Creation of a "Balancing" Atrial Septal Defect. While most operators do not perform this additional step we feel that it affords a great additional measure of safety While a special punch (Fig 24) 7 mm. in diameter has been designed especially for this purpose a suitable septal defect can be established with scissors under direct vision. However since the latter technic is an open one, it is diffi

cult to prevent entrance of air into the left atrium If the atrial septal defect is created before closure of the ventricular septal defect (but after performance of the right ventriculotomy) no special additional steps will be necessary to "vent" the left side of the heart of any entrapped air

A vertical incision made into the wall of

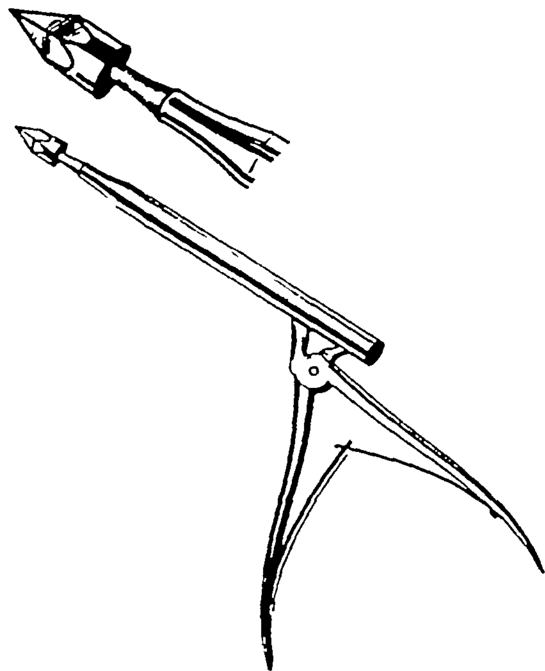


Fig 24 Punch designed for the creation of a "balancing" atrial septal defect of measured size

the right atrium will provide excellent exposure of the interatrial septum Using fine toothed forceps, and perhaps a sharpened *nerve hook,* an 8 to 10 mm linear incision is made in the thick muscular tissue of the dorsal portion of the septum just above the orifices of the right pulmonary veins (Fig 25) As soon as a sufficiently large opening has been established the lateral artial wall is closed in two continuous suture rows, one of mattress type and one a simple running stitch

Repair of Right Ventricular Wall The incision in the right ventricular wall is closed first with a continuous everting full thickness mattress suture preferably of moderate sized silk This suture line is begun close to the pulmonary arterial root and is continued perhaps with one or two interruptions to the opposite extremity of the wound Any bleeding ends of divided coronary arterial branches are picked up and ligated individually Just before closing the last two centimeters of the opening, the operator's index finger is inserted into the ventricular chamber to be certain that no intracardiac blockage or narrowing has been produced Before tightening the last few stitches, one of the vena caval tourniquets is released, permitting the right ventricle to become filled with blood, thus displacing any contained air

Termination of the Bypass Once the heart has resumed vigorous contractions and it has been determined that no element of

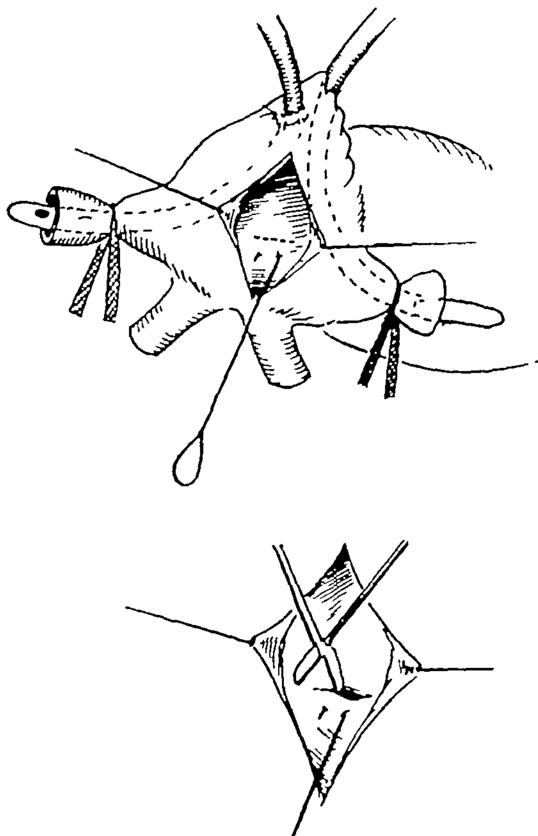


Fig 25 Creation of an atrial septal defect by incision of the right atrial wall, elevation of the muscular (dorsal) portion of the atrial septum, and linear incision by scalpel or scissors

heart block exists, both vena caval tourniquets are removed and the rate of bypass is slowed progressively to a level of from 200 to 500 cc of blood per minute If the mean blood pressure has not fallen during this very considerable reduction in the rate of perfusion, and if cardiac vigor seems unimpaired the pump is stopped and the catheters used for the vascular cannulation are clamped If the cardiac action and blood pressure remain satisfactory, *polybrene (arginine glutamate)*[51] is administered by intravenous drip in dosage equal to or slightly larger (1½ times) than that of the initially administered

heparin sulfate solution The heparinized blood used to prime the heart-lung apparatus must be considered in this calculation, and in larger patients is rated at one-half of its actual heparin content The polybrene is administered via intravenous drip over a ten minute period. A normal clotting time (Lee White technic) should be restored within 15 or 20 minutes after completion of the polybrene administration.

Decannulation: The superior vena caval catheter is clamped distally the purse-string suture about it is divided, and a noncrushing clamp is applied across the incised portion of the atrial wall as the catheter is withdrawn. The excluded portion of the right atrial appendage is tied off proximal to the clamp with a circular ligature of heavy suture material and the incisional opening is oversewn with fine arterial silk. The inferior vena caval (atrial) entrance site is treated in similar fashion

The femoral arterial cannulae are removed while the vessels are occluded temporarily both above and below the site of incision. The arterial incisions are repaired with fine interrupted arterial silk When the arterial cannula has been inserted through the anterior aspect of the ascending aorta the aortic wall is pinched together with a dentate clamp as the catheter is withdrawn. The aortic incision then is oversewn with a continuous suture of fine (5-0) silk.

The polyethylene tubes used for monitoring are removed and the respective vascular walls are repaired in simple fashion.

Chest Closure and Drainage During the period of progressive heparin neutralization, maximal hemostasis by all mechanical means (sutures, ligatures, electrocoagulation) is accomplished. The pericardium then is closed, leaving a large aperture at the inferior aspect of the incision for drainage. If either pleural cavity has been entered during the procedure, at this time the opening is repaired after an intercostal tube has been placed for water-seal drainage. Otherwise, the only drainage which we employ consists of a Lloyd double lumen multi-windowed catheter (Fig 26) which is inserted through the inferior extremity of the thoracic incision into the pericardial cavity Care must be taken to insure the placement of the double catheter tip beneath and behind the ventricles, and not in the recess between the right atrium and the lateral pericardial wall. The selected location insures maximum efficiency in lavaging and draining the pericardial sac.

Intermittent in-and-out pericardial lavage is carried out, beginning with the patient upon the operating table This is repeated at frequent intervals by the resident physician for hours (or days) until the wash solution returns entirely clear and all evidence of bleeding has ceased A satisfactory solution consists of a mixture of 10 cc of 1 per cent *neomycin* and 10,000 units of *bacitracin* in a liter of normal saline. The lavaging should be performed as necessary to keep the pericardium cleansed (usually every 5 to 15 minutes for the first few postoperative hours) under a physician's immediate supervision A

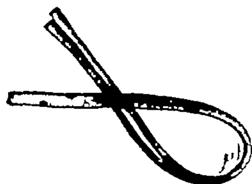


Fig. 26: Milton Sills Lloyd multi-windowed double catheter which has proven most advantageous for irrigation of the pericardial sac.

continuous unwatched drip must be avoided because there is a considerable danger of pericardial tamponade Usually this tube is removed on the second postoperative day the drainage tract being left unsutured to preclude the possibility of late development of tamponade.

Interrupted braided wire sutures are placed at 2 cm intervals to unite the sternal halves, using either cutting-edge needles or a punch to pierce the sternum. Heavy towel clips are useful for approximating the sternal edges as the wires are being tied The fascial and subcutaneous tissues superficial to the sternum, must be reapproximated carefully as subcutaneous sinus formation is an ever present hazard.

Special Postoperative Considerations

In addition to the customary postoperative complications such as hypo

volemia, atelectasis, etc., there are certain special hazards[52] to which one must be alert

The development of pericardial tamponade should be suspected if the patient shows any untoward sign suggestive of circulatory failure It can occur with amazing rapidity and without any of the usually accepted signs of its presence A sterile gloved finger inserted through the lower end of the incision into the pericardium and passed beneath the ventricles alongside the drainage tube will promptly rule out such tamponade or will confirm its presence and permit simultaneous correction by evacuation of the sac

Pneumothorax may occur even though the mediastinal pleura has been thought to be intact. This can be rapidly fatal, especially if bilateral, and must be promptly diagnosed and dealt with. Radiographic study performed while the patient is still on the operating table offers the best means establishing the presence of compl pulmonary expansion. However, dist or absent breath sounds in a reson chest afford presumptive evidence such a pneumothorax.

Complete heart block may occ immediately or several hours after s gery, presumably then being due to development of edema about the co mon conduction bundle. It is diffic to treat, and vigorous measures, inclu ing electrical myocardial stimulati with a pacemaker after the implan tion of wire electrodes may be need (Fig 27).

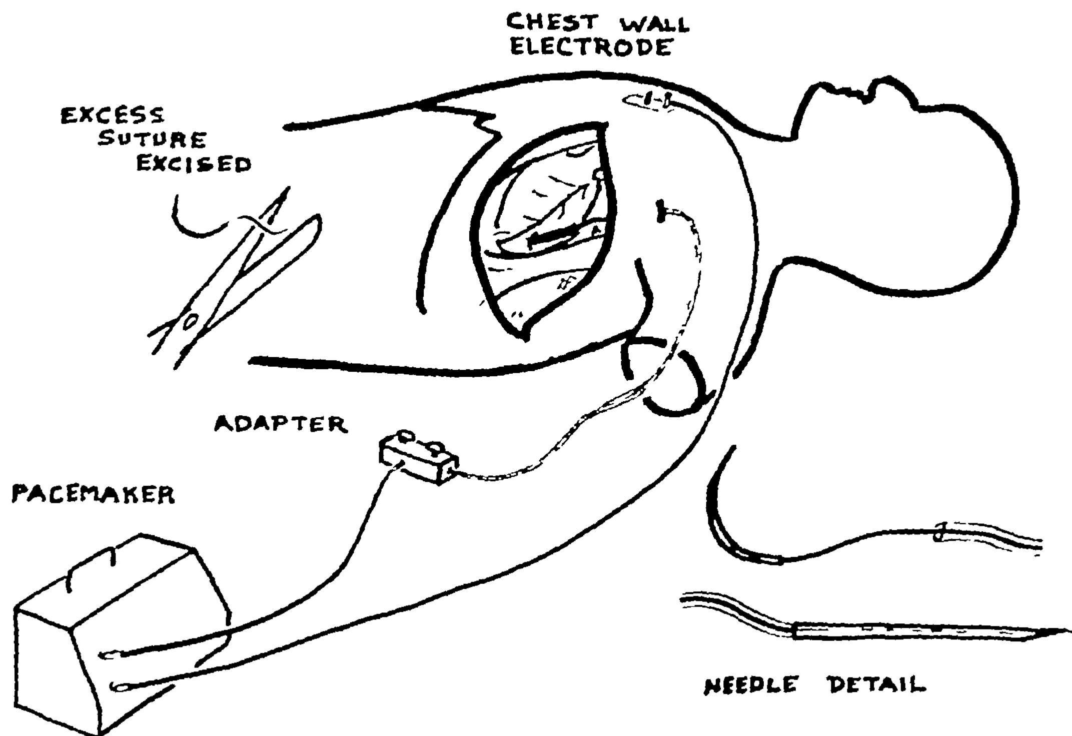


Fig 27: Method of stimulating ventricular contractions in heart-block by a pacemaker. Diagrammatic hook-up for pacemaker using both direct myocardial and indifferent electrodes

A moderate degree of hypervolen is not uncommon after cardiopulmona bypass, and must be relieved ev though the small peripheral vessels m prove difficult to utilize for venesc tion Prompt direct ventricular t under electrocardiographic monitori (if readily available) sometimes m

be necessary to abort an attack of acute pulmonary edema or congestive failure.

Myocardial irritability, evidenced by electrocardiographic abnormalities, must be reduced to prevent ventricular fibrillation. Currently at our clinic, continuous intravenous infusions of *xylocaine*, and parenteral *vistaril* are in favor

REFERENCES

1 Warden, H E, et al J Thoracic Surg., 33 21, 1957
2. Edwards, J E and Burchell, H B Thorax, 12 125 1957
3 Cooley D A Dis. Chest, 35 651, 1959
4 Lev M. L. Ibid., 35 533 1959
5 Kirklin, J W Harshbarger H G Donald, D E., and Edwards, J E. J Thoracic Surg., 33 46, 1957
6. Edwards, J E. Twenty-sixth Ludvig Hektoen lecture. Proc. Inst. Med. Chicago, 18 134 1950.
7 Edwards, J E. Henry Ford Hospital International Symposium on Cardiovascular Surgery W B Saunders Co Philadelphia, 1955
8 Eldridge, F L., Hultgren, H N and Wigmore, M. E. J Clin. Investig., 34 987 (July) 1955
9 Wood, E. H., Swan, H J C., and Marshall, H W.. Proc. Staff Meet. Mayo Clin., 33 536 (Oct. 29) 1958.
10 Baker R. R., Braunwald, E., Tanenbaum, H L and Morrow A G Ann. Surg 147 191 1958.
11 Sanders, R. J and Morrow A G Bull. Johns Hopkins Hosp 103 27 1958
12 Edwards, J E. Pediat. Clin. North America, pp. 13-49 (Feb) 1954
13 Shumacker H B Jr.. Angiology 5 289 1954
14 Kay E. B and Zimmerman, H A J.A M.A 154 986, 1954
15 Bailey C. P Surgery of the Heart. Lea & Febiger Philadelphia, 1955
16. Bigelow W G Hypothermia. Ann. Surg 43 683 1958.
17 Sellick, B A. Lancet, 272 443 1957
18 Wulff H B., et al.. Acta. chir scandinav., 112 186, 1957
19 Radigan, L. R., Lombardo, T A and Morrow A G S. Forum, 6 137 1955
20. Lillehei, C. W et al.. Dis. Chest, 29 1, 1956.
21 DeWall, R. A., et al J.A.M.A., 165: 1788, 1957
22 Gibbon, J H., Jr.. Minnesota Med., 37 171, 1954
23 Kirklin, J W., et al. Ann. Surg., 144 2, 1956.
24 Effler D B., Groves, L. K., Sones, F M., Jr., and Kolff W J Cleveland Clin Quart., 23 105 1956.
25 Kay E. B and Cross, F S.. Surg Gynec & Obst., 104 701, 1957
26. Grau, H R. Transpl. Bull. July and Oct., 1956 and Jan., Apr., and July 1957
27 Collins, H A., et al Surgery 45 506, 1959
28 Edwards, J E. M Clin. North America, 42 1037 1958
29 Muller W H., Jr and Damman, J T Ann. Surg 143 816, 1956
30 Therkelsen, F Boesen, I., and Vesterdal, J Minerva Cardioangiol. Europa, Tor., 4 224 1957
31 Sirak, H D., Hosier D M., and Clatworthy H W., Jr New England J Med., 260 147 1959
32 Kolff W J et al Cleveland Clin Quart., 25 65 1958
33 Sirak, H D and Hosier D M J Thoracic Surg., 37 1, 1959
34 Lillehei, C. W Ibid., 37 272, 1959
35 Benjamin, R. B Flom, R. S., MacLean, L. D., and Lewis, F J Ibid 34 679 1957
36 Wilson, H E., III Personal communication to C. P B.
37 Cooley D A., et al. Am. Surgeon, 24 870, 1958
38. Larios, R., Fitch, E. A., Blanco, G., and Bailey C. P: The Use of an artificial foraminal valve prosthesis in the closure of interatrial and interventricular septal defects. Presented at the 5th International Congress on diseases of the chest of the American College of Chest Physicians, Tokyo Sept. 7 1958 To be published in Dis. of Chest.
39 Bailey C. P Gould's Pathology of the Heart. Charles C. Thomas, Springfield, Ed 2. In press.
40. Bailey C. P., Zimmerman, J and Likoff W.. Dis. Chest. In press.
41 Allen, J G.. Extracorporeal Circulation. Charles C. Thomas, Springfield, 1958
42 Sealy W C., Brown, I. W., Jr., and Young W G Jr Ann Surg., 147 603 1958
43 Fitch, E. A. and Bailey C. P J Thoracic Surg 37 660, 1959
44 Zimmerman, H A., et al Ibid., 36 12, 1958.
45 Brechner V L., Kavan, E. M and Dillon, J B Ibid 37 352, 1959

46 Smith, W W, Brown, I W, Jr, and Eadie, G S A new anticoagulant-preservative solution for donor blood for use in extracorporeal circulation. Presented at San Francisco Meet Am Assoc for Thoracic Surgery, April 21-23, 1959 To be published

47 Satinsky, V P Personal communication to C P B

48 Zimmerman, J Personal communication to C P B

49 Helmsworth, J A., et al Ann Surg., 149 200, 1959

50 Truex, R C and Bishop, J K J Thoracic Surg 35 421, 1958

51 Weiss, W A, Gilman, J S, Catenacci, A J., and Osterberg, A E J A M.A., 166 603, 1958

52. Effler, D B, Kolff, W J, and Groves, L K Surgery, 45 149, 1959

SURGICAL TREATMENT OF TETRALOGY OF FALLOT

Charles P Bailey, M.D., and Edward A Fitch, M D

The complex of cardiac anomalies which combine to form the condition known generally as the tetralogy of Fallot[1 2] was recognized by Sandifort[3] in 1777 Contrary to Fallot s classical characterization of the pathophysiologic syndrome as comprised of over riding of the aorta, pulmonic stenosis, high ventricular septal defect, and hyper trophy of the right ventricle, it now is accepted by many that only two of the described anatomical features are essential for the typical clinical picture.[4] In the presence of a severe degree of pulmonic stenosis (which may reach a state of actual atresia) a coexisting ven tricular septal defect serves as an accessory outflow tract for the right ven tricle. Hence, there is a right to-left shunt at the ventricular level with resultant undersaturation of the systemic arterial blood. Commonly this is manifested by generalized bodily cy anosis which, when severe, usually is associated with clubbing of the fingers and toes The classically described "over riding' or "dextro-posed aortic root actually may exist. Frequently it is but an illusion caused by the absence of the upper oblique portion of the ventricular septum The very obvious hypertrophy of right ventricle is a purely compensatory development which enables this chamber to compete functionally with the left ventricle.

It long has been recognized that stages of this malformation may exist which are less severe clinically [5 6] Thus, the ventricular septal defect which usually is very large may be diminutive or the usually severe pul monic stenosis may be mild. Then the tendency toward right to-left shunting through the septal defect may be small. Indeed, in some cases the tendency toward left to-right shunting through the ventricular septal defect may be exactly equal to this so that no shunt ing or but small equalized increments of biphasic shunting will take place Physiologically speaking one then might conceive the defect to be non functional although dynamic competi tion between the two communicating ventricles necessarily must exist. At any rate in these cases little or no under saturation of the arterial blood will be found. These patients have been de scribed as having acyanotic tetralogy of Fallot."

Even less pronounced degrees of pulmonic obstruction may exist so that the condition assumes more and more the characteristics of an isolated ven tricular septal defect. Then a signifi cant left to right shunt may exist These cases also have been categorized as representative of *acyanotic tetral ogy* [7 8] If the shunt is large pulmonary hypertension and degenerative pulmonary vascular changes such as occur in isolated ventricular septal defects may develop Thus, one may visualize a smooth spectrum of clinical and phy siologic variances, depending upon the magnitude of the pulmonic obstruction and the septal defect, ranging from a state of true pulmonic atresia with total right to-left shunting through the septal defect to that of an isolated ven tricular septal defect with a left to-right shunt. Again, the defect may range from huge to such insignificance that the condition may approach the state of "pure" or isolated pulmonic stenosis (Fig 1A,B C,D E)

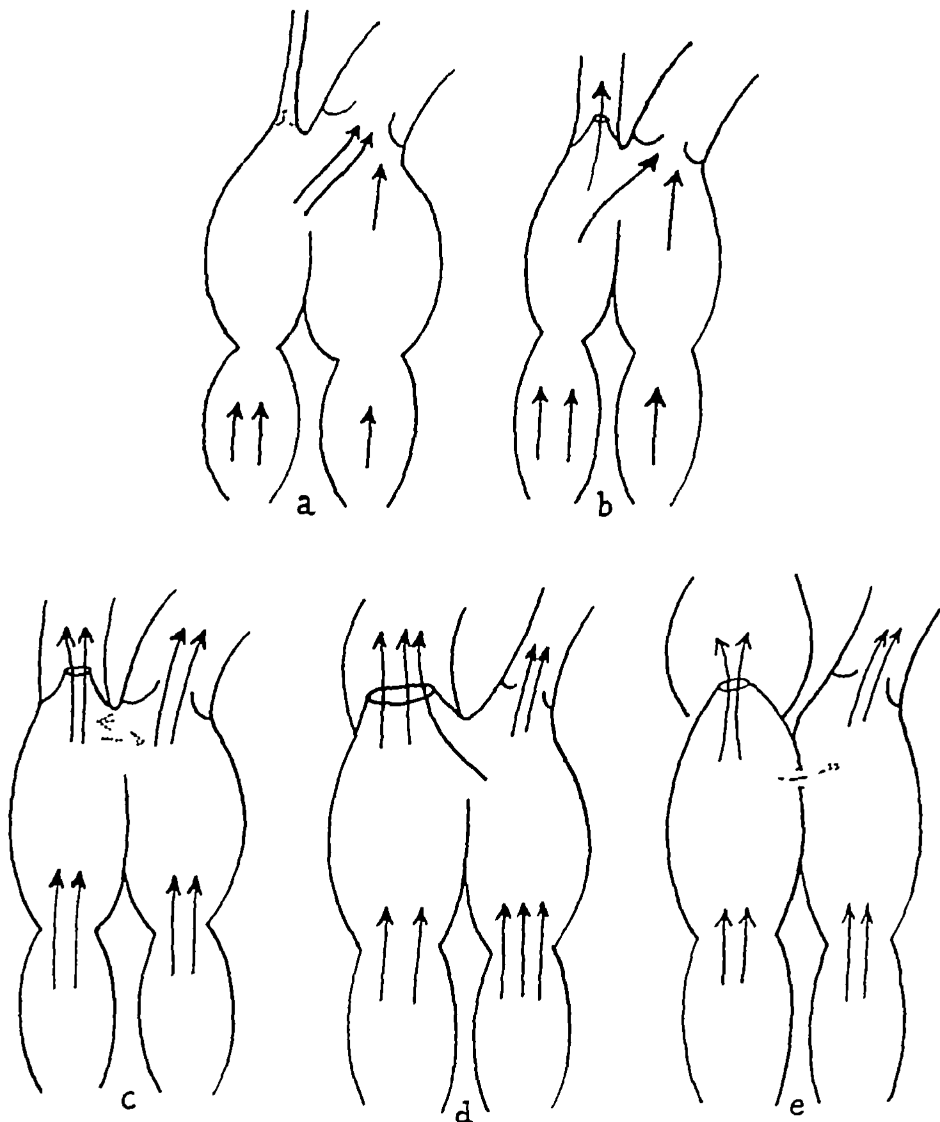


Fig 1 Functional variation in the complex known as the tetralogy of Fallot *a*, Extreme pulmonary stenosis approaching or attaining the state of complete pulmonary atresia Practically all of the right ventricular blood content passes through the septal defect to enter the aorta Life is maintained by collateral mechanisms *b*, Classical variety of the tetralogy of Fallot Due to severe pulmonary stenosis (valvular, infundibular, or both) a large portion (but not all) of the right ventricular blood passes through the defect to enter the aorta (right to left shunt) *c*, "Acyanotic Tetralogy" Because the pulmonary stenosis is merely of moderate severity the tendency toward right-to-left shunting is exactly balanced by the tendency toward left to right shunting through the defect A small amount of equalized biphasic shunting may be present *d*, When the pulmonary stenosis is slight, a left-to right shunt occurs through the defect so that physiologically, the case resembles one of isolated ventricular septal defect *e*, If the defect is extremely small so that it is nearly nonfunctional, the case resembles one of isolated or "pure" pulmonary stenosis

Pathologic Anatomy

The severity of the impedance to flow through the pulmonary artery varies greatly. This is a function of the type and severity of the specific anatomical deviation In some cases an hypertrophied supraventricular crest may be found in the presence of a large well marked infundibular chamber (Fig 2A) In others the crest may lie close to the pulmonary valve so that but a small infundibular chamber is present (Fig. 2B). A hypoplastic and generally constricted right ventricular outflow tract may exist, frequently in association with a state of hypoplasia or atresia of the pulmonary artery (Fig 2C)

At times the cause of the obstruction may be a congenital fusion of the pulmonary valve whether it be of tricuspid or bicuspid structure (Fig 2D) In the majority of clinically severe cases of the tetralogy of Fallot the pulmonary artery is much smaller than normal, sometimes, as mentioned, being reduced actually to the point of atresia. Not infrequently more than one site or type of pulmonic stenosis may exist.

The septal deficiency may assume

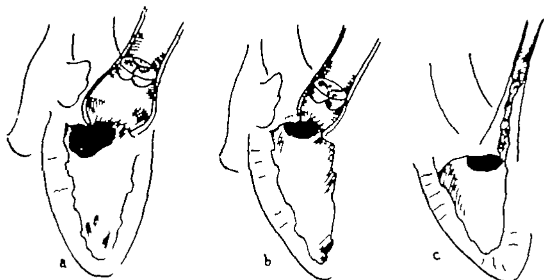


Fig 2: Anatomical variations in the complex known as the tetralogy of Fallot. a, Low infundibular obstruction with formation of a good sized infundibular chamber b High infundibular obstruction with formation of a small infundibular chamber c, Extreme hypoplasia of the right ventricular outflow tract Usually the pulmonary valve and main arterial trunk are involved in the hypoplasia.



Fig 2: D Photograph of a stenotic pulmonary valve (congenital) showing obliteration of all three commissures by symmetrical fusion of the cusps.

any of the forms and localizations described in Ventricular Septal Defect. In general, however the septal defect in the tetralogy of Fallot tends to be rather large and presents itself in close relation to the cusps of the aortic valve so that anatomically and physiologically it can serve as an additional or auxiliary right ventricular outflow tract.

Clinical Picture

As stated, in the more severe states of the tetralogy of Fallot the patient exhibits generalized cyanosis and clubbing of the fingers and toes. This evident cyanosis is a resultant both of the undersaturation of the arterial blood and the rather marked polycythemia which usually develops. Physical exertion, even when of a minimal type, causes an increase in the under

saturation of the arterial blood, producing shortness of breath and weakness or dizziness Commonly, these symptoms are relieved by *squatting* or assuming the *knee-chest position* (Fig. 3) In the more severe cases in which the level of arterial oxygen saturation is very low,



Fig 3 Typical "squatting" position often assumed by patients with the tetralogy of Fallot when physical activity has exceeded the functional limits of the circulation (Bailey, C P Surgery of the Heart Lea & Febiger, Philadelphia, 1955)

episodes of convulsions and unconsciousness may follow the slightest physical effort. In the most severe cases it may be impossible to remove the infant from an oxygen enriched atmosphere for more than a few minutes at a time Life expectancy in such individuals may be measured in days or weeks In clinically less severe cases life may be maintained for months or years, but necessarily all physical activities must be restricted, often to an intolerable degree Death in early childhood is the rule although some individuals, even some of those with severe grades of pulmonic stenosis and marked undersaturation of the arterial blood may survive into adult life. These patients present a picture of marked cyanosis, chronic reddening of all mucosal surfaces and especially of the conjunctivae, extreme clubbing of the fingers and toes, and, in many instances, a marked deficit in the mechanism of blood coagulation. Their survival may be attributed to the presence of an excessively overdeveloped collateral pulmonary vascular system which is based chiefly upon the ramifications of the bronchial arteries In some cases left-to-right shunting through a patent ductus arteriosus may provide partial but reasonably effective compensation for the right-to-left shunting through the ventricular septal defect As part of the universal dilatation of the systemic arteriolar and capillary beds an increment of compensation is offered by numerous small mediastinal and thoracic wall vessels which enter the pulmonary hilum or reach the lung through pleural adhesions either of congenital or acquired origin Indeed, Barrett[9] has recommended the surgical creation of pleural adhesions in patients with actual pulmonary atresia or extreme pulmonic stenosis in order to enhance this mechanism These minute systemic arteries communicate with the pulmonary capillary bed and therefore conduct a modicum of undersaturated arterial blood to the pulmonary alveoli Oxygenation of this mixed blood then takes place in the usual manner

It must not be overlooked that patients with less severe grades of pulmonic stenosis may do very well clinically manifesting little or no cyanosis,

except perhaps, upon the most vigorous exertion (*acyanotic tetralogy*) However even in the absence of significant right-to-left shunting after the patient has reached a certain age (30 to 40 years) symptoms tend to appear which are attributable to a low cardiac reserve. Eventually frank heart failure develops. Undoubtedly the anatomically very differently constructed right ventricle[10] cannot compete indefinitely with the architecturally more powerful left ventricle with which it has been in direct dynamic continuity and competition over this prolonged period milder cases the vascular shadows often are quite within normal limits (Fig 5B) In extremely polycythemic individuals the lung fields may show many streaks of *fibrosis* presumably related to intravascular thrombosis or to resolution of areas of old infarction.

The *pulmonary* or *conus segment* of the cardiac silhouette commonly presents a concave rather than the usual convex outline. The apex of the heart not infrequently is elevated due to the right ventricular hypertrophy This along with the concave *conus segment* may present an appearance which has

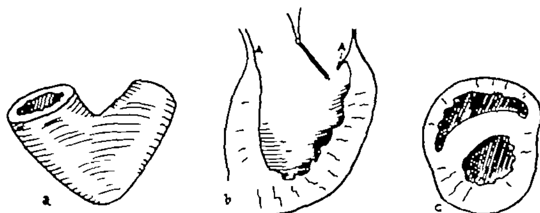


Fig 4 Structural differences between the right and left ventricles. a The right ventricle is a flattened hollow tube which contracts in "peristaltic" fashion. b, The left ventricle is a hollowed-out muscular cone which in effect has but one aperture of ingress and egress. c, View which shows cross-sectional outlines of the two ventricles. The right is applied about the left in such a manner as to be partially "wrapped around" the latter

(Fig 4A,B C) Presumably a similar fate awaits those whose clinical condition has been improved by a Blalock-Taussig or Potts-Smith shunt,[11 14] or even by a Brock type of subtotal relief of the pulmonic obstruction,[15 18] unless definitive correction of the fundamental lesions can be carried out subsequently

Radiographic Features

Characteristically the peripheral lung fields in the tetralogy of Fallot are said to be *clear* due to the presence of less than usually prominent vascular markings (Fig 5A) However the hilar vessels may be of fair size, and in been described as that of the *Dutch Boot* (Fig 5A)

Angiocardiography: Angiocardiography by the venous route usually reveals simultaneous opacification of both the aorta and the pulmonary vessels (Fig 6A,B) In cases with atresia or near atresia of the pulmonary artery while the aorta fills early very late, slight, or no opacification of the lesser circulatory bed may be recognized.

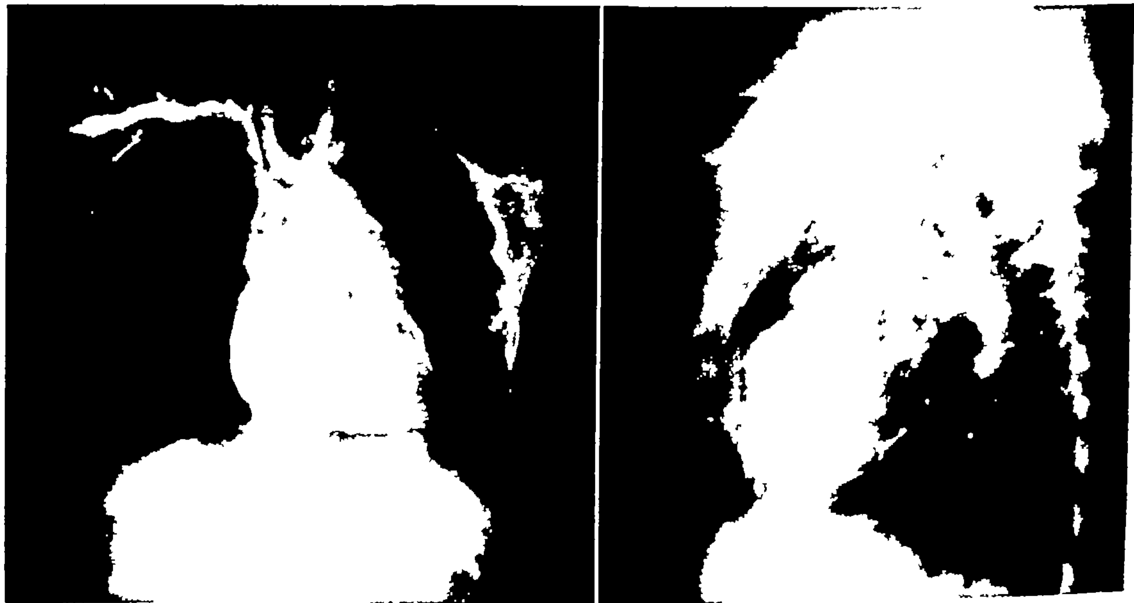
Physical Signs

In addition to cyanosis at least of the nail beds, and digital clubbing (when present) there often is generalized



A B

Fig 5 Radiologic appearance in the tetralogy of Fallot A, Typical "Dutch Boot" cardiac outline often seen in severe states of the tetralogy of Fallot Note the relatively transparent lung fields due to the diminutive size of their vascular components B, Relatively normal radio graphic configuration of the heart which may be seen in the tetralogy of Fallot particularly in less severe states The pulmonary vascular markings are within normal limits (Bailey, C P Surgery of the Heart Lea & Febiger, Philadelphia, 1955)



A B

Fig 6 Simultaneous opacification of the aorta and pulmonary artery by venous angio cardiography This finding establishes both the presence of a ventricular septal defect and the existence of a right to left shunt A, Posteroanterior view B, Lateral view (Bailey, C P Surgery of the Heart Lea & Febiger, Philadelphia, 1955)

congestion of all mucosal vessels (notably those of the conjunctivae) The usual auscultatory finding is a loud harsh systolic murmur maximal over the midsternal region, often associated with a palpable thrill However in the presence of complete pulmonary obstruction (atresia) no murmur may be audible. Characteristically the pulmonary second sound is faint or absent In "acyanotic tetralogy" the systolic murmur may be slight or absent presumably because of the 'equalization or "balancing" of the circulation.

Catheterization Findings

The measured systolic pressure within the right ventricle will be the same as that within the left ventricle and the systemic arteries, but will be much higher than that in the pulmonary artery When an infundibular chamber exists a site of intermediate pressure will be encountered (Fig 7) When there is only a right-to-left shunt there is no step up in the oxygen saturation of blood specimens obtained at the ventricular level. In cases of acyanotic tetralogy' the catheterization findings usually are similar to those of an isolated ventricular septal defect except, of course, that the systolic pulmonary arterial pressure is significantly lower than that within the right ventricle Often in the tetralogy of Fallot the catheter tip will not enter the obstructed pulmonary circulation but will pass (repeatedly) through the ventricular septal defect to enter the aorta. In all but the cases of *acyanotic tetralogy* the brachial arterial oxygen saturation will be significantly lower than normal (Tables 1 and 2)

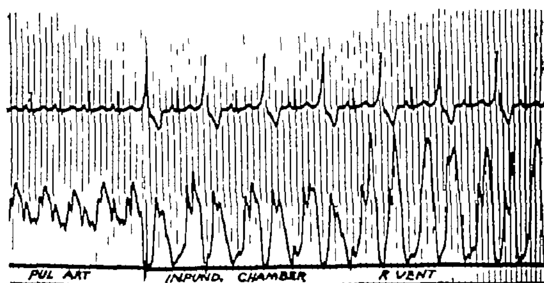


Fig 7 Demonstratio of the presence of infundibular stenosis and a well ma ked infundibular chamber during the "pull-back" of the cardiac catheter tip from the pulmonary artery (lower left tracing) into an infundibular (ventricular) chamber (lower central tracing) and finally into a higher pressured ventricular chamber (lower right tracing). Note the alternation of normal and bnormal ventricular contractions (electrocardiographically) which bega as the catheter tip entered the ventricle.

Diagnosis

If the catheter tip can be caused to enter the pulmonary artery the lower pressure within this chamber in contrast to that within the right ventricle (which nearly always is equal to that within the left ventricle) will establish the existence of pulmonic stenosis.

TABLE 1

Cyanotic Tetralogy — Case Y. P.*
Oxygen Content

	Preoperative	*Postoperative*
SVC	16.2	13 1
RA	17 4	11 9
IVC	14.2	
TRICUSP	16 7	11 9
RV	18 7	13 1
PA	14 2	12
RPA	16.2	

TABLE 2

Acyanotic Tetralogy — Case P. J.*
Oxygen Content

	Preoperative	*Postoperative*
SVC	14 6	9 6
RA	14 7	9 2
IVC	13.2	
TRICUSP	14 1	9.5
RV	14 1	10 2
PA	13 2	10 1
RPA	13 4	10

* The lower oxygen content of blood specimens postoperatively is related to abolition of the polycythemia

Often an area of intermediate pressures will be encountered which indicates the anatomical presence of an infundibular chamber (Fig 7) This establishes the presence of infundibular obstruction but does not disprove the presence of coexisting pulmonary valvular stenosis If the latter type of stenosis exists in the absence of an infundibular obstruction the pressure change will be abrupt and at the valve level More recent refinements in the technics of contrast radiography may enable one to demonstrate both the type and the severity of the pulmonic obstruction. Simultaneous opacification of the aorta and pulmonary artery during venous angiocardiography or the passage of the catheter tip from the right ventricle into the aorta will confirm the presence of a ventricular septal defect For clinical purposes the combination of pulmonic stenosis with a ventricular septal defect is considered sufficient to establish the presence of the tetralogy of Fallot.

When the catheter fails to enter the pulmonary circuit or the presence of a ventricular septal defect is not demonstrated incontrovertibly, one may have to rely upon such indirect evidence for the diagnosis as generalized cyanosis in the presence of a small heart with a suggestive radiographic outline and the lack of normally intense pulmonary vascular markings (Fig 5) Equalization of the pressures within the two ventricles suggests the presence of a large communication (septal defect) The presence of a state of "acyanotic tetralogy" cannot be diagnosed with confidence without catheterization proof of the existence of a degree of pulmonic stenosis plus a ventricular septal defect and either predominant left-to-right or biphasic shunting

Surgical Treatment

The surgical treatment of the tetralogy of Fallot may be divided into three types or methods (1) palliative, (2) physiologic, and (3) corrective

The *palliative* treatment includes the various technics for creation of a systemic-pulmonary arterial anastomosis whether it be between the subclavian artery and the pulmonary artery (Blalock-Taussig[11]), or between the descending aorta and a pulmonary artery (Potts-Smith[12]) or some other technically feasible anatomical variant. In any event a left-to-right shunt is established through an artificially created "patent ductus arteriosus" This tends, in large measure, to counter or overcome the effects upon the circulation of the right-to-left shunt at the ventricular level. Expressed differ-

ently a portion of the mixed saturated and unsaturated blood within the systemic arterial system is caused to return to the pulmonary artery whence it passes through the lungs for further oxygenation. While these procedures have the effect of adding still another circulatory anomaly to those already extant, they have sufficient compensatory value to lessen the over all severity of the clinical picture considerably However since nothing will have been done to alter the intrinsic intracardiac malformations the right ventricle must remain in dynamic competition with the left, a burden which it scarcely can be expected to maintain indefinitely [13 14] Furthermore, should too large a shunt be established the cardiac competence will be compromised immediately This may lead to an early onset of signs of congestive heart failure.

Nevertheless, the creation of such an artificial patent ductus arteriosus still holds an important place in the treatment of certain types of patients such as those with actual pulmonary arterial atresia and also infants who may be too feeble to tolerate a fully corrective operative procedure. In this latter instance the palliative operation probably will enable the individual to survive until he will have become large and strong enough to accept a more major fully corrective intervention.

The *physiologic* approach to severe states of the tetralogy of Fallot consists of an attempt to reduce the magnitude of the right to-left shunt to insignificance by diminishing the severity of the pulmonic obstruction by pulmonary valvulotomy [15 16] infundibulectomy [17] enlargement of the right ventricular outflow tract,[18] or some other suitable procedure. Actually these efforts amount to attempts to establish a state of 'acyanotic tetralogy" in which the tendency toward a right-to-left shunt through the defect will be balanced exactly by the tendency toward a left to-right shunt which develops in response to the surgical reduction in the severity of the pulmonic stenosis. Thus, one may hope to overcome all of the immediately serious physiologic effects of the malformation Of course the two ventricles necessarily will continue to remain in dynamic functional competition and ultimate failure of the right ventricle might well be predicted in the very distant future.[19 20]

Therefore, the profession is inclined to relegate the *physiologic* operations also to a status of temporizing or preparatory procedures, which presumably are to be followed at a more favorable time by more definitive fully corrective surgery The usually rather diminutive outflow tract and the hypoplastic pulmonary artery tend gradually to become enlarged by the increased pulmonary blood flow established by these technics. Furthermore, the relatively underdeveloped pulmonary arteriolar and capillary beds which initially may be quite incapable of transmitting the entire circulatory flow at normal pressures progressively become larger In time they will become dilated sufficiently to accept not only the entire right ventricular output but perhaps even a significant additional increment of flow which will be added if a left-to right shunt develops.

While two-stage surgery for the tetralogy of Fallot carries with it certain undesirable implications (repeated operative jeopardy the necessity at the second stage for dealing with vascularized intrapericardial adhesions, and the obliteration of normal surgical landmarks) it may represent the only

feasible operative approach[21] in certain individuals, such as those presenting extreme cyanosis and marked hypoplasia or actual atresia of the outflow tract, or extreme youth (infancy).

In the past, it has been necessary that pulmonary valvulotomy, infundibulectomy, or dilatation of the pulmonary artery be performed by a closed operative technic. Such surgery may prove life-saving and often has seemed gratifyingly effective However, since these are essentially "blind" procedures, and since there may be infinite variation in the details of the pathologic anatomy, at times little effective enlargement of the site of maximal obstruction has been accomplished by such technics. At other times so much of the pulmonary blockade has been removed that a very large left-to-right shunt has been created In some of these patients effective compensation has not been possible. Blind pulmonary valvulotomy rarely can fail to produce pulmonary incompetence, if an adequate opening is provided (Fig 8A,B,C). In a few instances during the blind performance of infundibular resection serious damage has been done to the aortic valve (partial avulsion of an aortic cusp, perforation of a sinus of Valsalva) so that aortic incompetence has ensued (Fig. 9A,B,C). The over-all operative mortality of these blind procedures for modification of the clinical state in the tetralogy of Fallot has been reported at 14 per cent by Brock.[22]

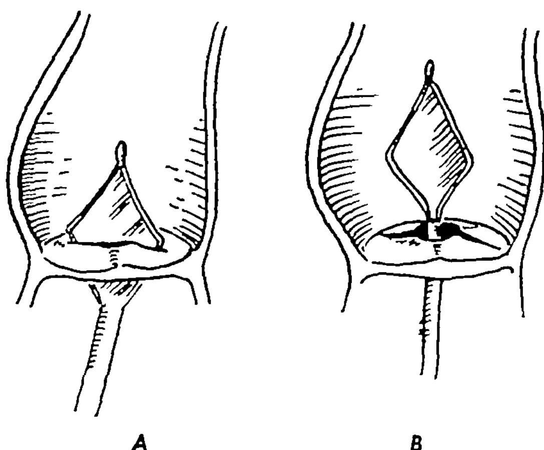


Fig 8 Closed valvulotomy for pulmonary valvular stenosis (Brock technic) This technic nearly always causes a significant amount of incompetence *A*, Advancement of the diamond-shaped valvulotome through the cap-like valve *B*, Logically the resultant line of incision may divide one commissure accurately while interrupting the continuity of the opposite cusp



Fig 8 *C*, Actual postoperative appearance of a stenotic pulmonary valve after blind valvulotomy In this case only a single radiating section is apparent Purely fortuitously a commissure was divided properly

For these several reasons, except in frank emergencies, it is our present feeling that the *physiologic* type of operation should be carried out only under direct vision Complete circulatory bypass with a pump-oxygenator, is essential for such a planned methodical procedure However, extensive development of the various elements of the collateral system, especially those which are based upon the bronchial circulation, is characteristic of severe cases of the tetralogy of Fallot Serious hemorrhage (from the pulmonary veins) may result once the heart is opened since the systemic arteries (in-

cluding the bronchial tributaries) continue to be perfused (by the pump) This excessive bleeding during operation may be avoided by the additional employment of general bodily hypothermia which permits use of low perfusion rates and low arterial pressures or even temporary circulatory arrest. The hypothermic state may be established most conveniently by incorporation of a heat-exchanger within the extracorporeal circuit as recommended by Sealy Brown, and Young [23] Drew and Anderson[24, 25] recently have recommended the use of extreme hypothermia (from 50 to 59 F [10 to 15 C.]) and been able in the more severe stages of the tetralogy of Fallot to prevent excessive bleeding from the opened heart in the presence of an extensive collateral system. In cases of "acyanotic tetralogy" on the other hand, conventional bypass at normothermic temperatures is satisfactory

The definitive operative steps necessarily include the establishment of the largest feasible right ventricular outflow passageway by infundibulectomy pulmonary valvulotomy, or by certain of several improvised or combination procedures which can achieve this end. It must be admitted at the outset, that



Fig 9 The essential (and usual) pathology of the tetralogy of Fallot comprises a high ventricular septal defect and a muscular ridge (supra ventricular crest) which approaches the anterior wall of the outflow portion of the right ventricle and tends to approximate its interior surface during systole View from below *a* Diastole—showing a fair-sized passage *b* Systole—showing increased obstruction which occurs during the period of ejection. *c*, Blind infundibulectomy by removing a portion of the ridge increases the size of the outflow passage during systole Note proximity of the aortic valve cusps.

prolonged interruption of the circulation in combination with resuscitative circulatory bypass.

The fully *corrective* approach in this condition implies a rather intricate and prolonged open operative procedure which can be performed only with complete cardiopulmonary bypass.[7,20,26] As discussed, it is eminently desirable also to establish a state of general bodily hypothermia so that only very low perfusion flows will be required. Controlled periods of complete interruption of the circulation will be permissible during this hypothermic state. Only by such a combination of methods have we establishment of an outflow passage of fully normal size is not always possible. Diminutive size of the pulmonary artery and/or valve, or marked hypoplasia of the infundibular region may preclude the attainment of an ideal state of correction. However for sedentary living and even for a life of considerable activity fully normal sized pulmonary arterial channels are not absolutely essential. If unavoidable the right ventricle can accept permanently a moderate degree of pulmonic stenosis providing the full effect of the hydrodynamic impedance is not imposed upon it too suddenly

The ventricular septal defect, however, must be closed, either by direct suturing, or more often, since the defects in this condition tend to be rather large, by the use of a prosthetic patch of plastic material The tendency for pure ivalon sponge when so utilized to become fragmented with the passage of time[27, 28] strongly commends the use of a permanently stronger material Teflon netting incorporated within a "sandwich" of compressed ivalon, as suggested by Larios and others[29] has proven to be a satisfactory and convenient material In general, the technics employed in closing isolated ventricular septal defects may be utilized for the correction of the defects associated with the tetralogy of Fallot Because of the large size of the transseptal communication it is especially easy to apply the sutures into the septal tissue from within the left ventricle in the region in which the common conduction bundle may be encountered

Prevention of Sudden and Severe Intracardiac Pressure Changes: If the atrial septum is intact and the defective ventricular septum is rendered so, all of the right ventricular outflow (*i e*, the full bodily venous return) thereafter must pass through the pulmonary artery and its tributaries Providing these passages are of adequate size, and the total pulmonary vascular resistance is not too great, this is eminently desirable since it permits proper oxygenation of the entire circulatory flow.

Since it is not always possible fully to correct the obstruction within the right ventricular outflow tract and pulmonary trunk, and since, especially in very cyanotic individuals, the minute pulmonary vascular bed may not be sufficiently developed to accept the entire cardiac output at once, immediate complete closure of the ventricular septal defect suddenly may impose an intolerable burden upon the right ventricle It may fail immediately or within the course of a few hours. Or, alternatively, extravasation of blood into the pulmonary parenchyma may occur[30] presumably due to leakage from the overdistended pulmonary capillaries This produces a liver-like condition of the lungs and in itself can precipitate respiratory failure

These two factors, along with other risks which are inherent in circulatory bypass for repair of defects of the ventricular septum in cyanotic patients have been responsible for an operative mortality for fully corrective procedures in the tetralogy of Fallot which at times has seemed prohibitive[7, 20, 31] This operative risk does not hold true in patients who are but slightly cyanotic or frankly acyanotic since these latter seem to be the most favorable of all patients with ventricular septal defects for definitive surgical correction Indeed some very able cardiac surgeons have accepted the dictum that "blue" tetrads must be operated in two stages (first a shunt or Brock type of procedure) while the milder "pink" tetrads may be corrected fully at one stage[21, 31]

It has seemed to the authors that the surgical risks associated with fully corrective operations for the tetralogy of Fallot often might be reduced in another way, although it is conceded that the two-stage concept should be honored in certain types of cases, notably in the very small infant and in the individual with a very tiny pulmonary artery and/or outflow tract Since it usually is impossible to relieve both the central and the peripheral (arteriolar and capillary) obstructions in the lesser circuit completely at one operative stage, it would seem eminently desirable

that the postoperative burden of propelling the entire circulating blood volume through this partially obstructed vascular bed should be imposed upon the right ventricle in as gradual a manner as possible. This might be accomplished in one of two ways. Either one might create a small gradually closing atrial septal defect such as has been utilized in order to equalize or balance the intra atrial

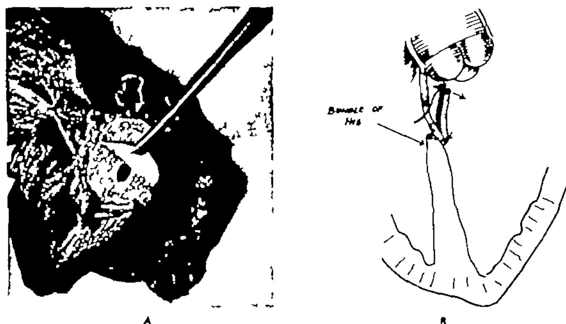


Fig. 10: The prosthetic foraminal valve is prepared at the time of surgery from a two-leaved patch of compressed ivalon sponge reinforced with teflo netting which is incorporated i "sandwich" fashio between layers of the spong A, Photograph showing such a prosthetic foraminal valve which had bee applied for the gradual closure of a very primitive transseptal communicatio (atrioventricularis comm nis) B Illustration showing the necessity for "tailoring" the flap of the prosthetic foraminal valve (applied in the tetralogy of Fallot) in such a way that it does no "rub ag inst" or distort the contiguous aortic valve cusps.

pressures in certain other conditions (as in correction of ventricular septal defects and in some of the modern operations for mitral stenosis)[32] or a gradually closing type of prosthetic device might be applied to the ventricular septal defect.[33, 34] Sirak and Hosier[35] have suggested the creation of an artificial patent ductus arteriosus at the time of closure of ventricular septal defects.

Extensive experimentation with the accomplishment of gradual closure of artificially created septal defects using prosthetic valves in animals, and early trial in clinical cases has convinced us that this approach is feasible. Of the two methods (creation of a systemic pulmonary circulatory shunt versus insertion of a prosthetic valve) the use of a slowly closing prosthetic valve for the ventricular septal defect has seemed to us the more logical procedure in the tetralogy of Fallot. The prosthetic artificial foraminal valve has been developed as the result of these investigations.[29] It consists of a slab of teflon reenforced compressed *ivalon sponge* which has been fashioned as two leaves which are joined securely at one quadrant. By cutting a central hole in one of the leaves a unidirectional flap valve with a lid and ring may be created (Fig 10A,B) By applying this prosthetic valve appropriately across the

ventricular septal defect the flap may be caused to open into the left ventricle thus permitting continuance of a right-to-left shunt (Table 3) The prosthetic valve tends inexorably to become sealed shut within eight to twelve weeks by virtue of envelopment and impregnation of each of its "porous" leaves with sticky fibrinous exudate. This exudate tends gradually to agglutinate the respective leaves together, and with organization (by fibroblastic invasion) eventually forms a tight fibrous bond Since the pressure within the right ventricle tends immediately or gradually to fall below that within the left (after the definitive correction of the basic deformities of the tetralogy of Fallot), it would seem that a prosthetic foraminal valve so placed as to permit a temporary maintenance of the right-to-left shunt should manifest an overwhelming tendency gradually to become closed. Cardiac catheterization after the elapse of some months has confirmed this repeatedly in clinical cases

TABLE 3

	Pre-Op	*Early Post-Op*
SVC	12 7	15.2
IVC	12 4	15
RA (High)	13 4	
RA (Mid)	13 1	15 4
RA (Low)	13 2	
RV (Tricuspid)	13 1	15.2
RV (Apex)	13 2	
BA Content	18 7	17 9
Capacity	20 I	21 1
Sat	87%	85%
Press	$\frac{120}{82}$ mm Hg	$\frac{128}{85}$ mm Hg
Mean	98	102
RV (Mean)	96 mm Hg	$\frac{115}{4}$ mm Hg

In the majority of patients v tetralogy of Fallot in whom tl ventricular outflow tract is ı tremely hypoplastic, and in wh to normal sized pulmonary vessels exist it seems to us that operative approach is eminently In practice it has proved to t satisfactory. In those patients w sent a very small or atretic ven outflow tract or pulmonary arte our feeling that a two-stage pr should be employed, not only vage the individual initially, b sibly also to "prepare" him for ary "curative" surgery by enco the development and enlargen the infundibular region and pul artery In general, we prefer to p a Brock type of procedure, u open technic (with circulatory since direct vision permits a co amount of enlargement of the channel.

Operative Technic

No description will be given the thoroughly established op technics for the performance Blalock-Taussig or Potts-Smith systemic arterial-pulmonary arte astomosis. These technics ar documented elsewhere[11, 12, 36, 3

It is felt that the closed techr the performance of pulmonary lotomy (Sellors-Brock) or parti sion of an hypertrophied suprave lar crest (Brock infundibulec likewise have been documente quately.[15, 16, 17] It is urged, hc that these basically blind proc except perhaps for the solutio near-emergency situation, shou replaced whenever possible by priate technics performed under vision control Necessarily this v quire either hypothermia or car

monary bypass, or preferably a combination of both. Drew and Anderson[24, 25] have utilized the combination of both a heat-exchanger and Blanco's[38] method of using the "autogenous lung" with circulatory bypass in order to establish an extremely low level of general bodily hypothermia, 50 to 59

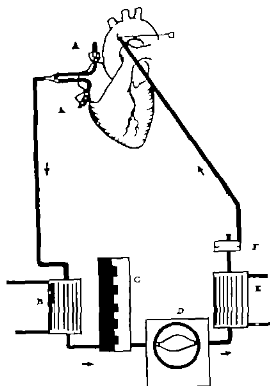


Fig 11 Recommended extracorporeal perfusion circuit for definitive correction of the pathology in the tetralogy of Fallot. A, Individual plastic catheters which have been inserted into the respective venae cavae. B Heat exchanger in the venous line (gravity drainage) for rewarming the patient. C, Oxygenator D Pump (preferably pulsatile). E, Heat exchanger in the arterial line for cooling F Bubble and fibrin filter trap. G Arterial catheter of large bore preferably inserted directly into the ascending aorta

F (10 to 15 C.) At such temperatures periods of complete circulatory interruption for as long as 45 minutes are apparently permissible. Usually we have preferred merely to reduce the temperature to such a level (68 F [20 C.]) that very low circulatory flow rates are required However appreciable periods of circulatory interruption still are permissible in the event that troublesome bleeding related to the overdeveloped bronchial collateral circulation is encountered. Combined cooling and bypass of this type at our clinic usually entails the incorporation of an artificial oxygenator as well as two heat exchangers within the extracorporeal circuit.

Cannulation for Bypass

The perfusion circuit, illustrated in Fig 11 is set up and the reservoirs and tubing are filled with fresh heparinized blood.

After a state of incoagulability of the circulating blood has been established by the administration of 3.0 mg *Heparin* per kg of body weight the multifenestrated tips of plastic catheters of appropriate size stiffened by insertion of Frech malleable obturators (Fig 12A,B) are advanced through individual purse stringed incisions made in the right atrial wall into the superior and inferior vena cava respectively A second (left) superior vena cava is sought and if found, either may be cannulated separately or simply encircled with a tape for constriction. Care is taken that all the fenestrations of the catheters lie well beyond the tapes which encircle the venae cavae since improper placement can lead either to massive bleeding or to air embolism. The purse string sutures are tied tightly are passed twice about the plastic catheters, and then are retied thus affixing them securely to the atrial wall After removal of the obturators, the catheters are attached to the "venous" lines of the extracorporeal system. If a bubble type oxygenator is used no great care need be taken to express all air bubbles from the tube junctions. With certain other types of oxygenators all air should be excluded.

The arterial cannulation may be performed in the course of the common femoral artery (or preferably both femoral arteries) in larger children or adults. In performing circulatory bypass in small children and infants (and even in adults) we much prefer to cannulate the ascending aorta at a site which lies well above the valve.[39] This may be carried out readily by encircling widely a conveniently located point upon the aortic wall with 2-0 nylon suture material swaged upon fine curved needles. It is preferable that the needle should not actually penetrate

the arterial lumen but should only "pick-up" the deepest layer of the adventitia The encircled area is excluded from the main aortic lumen by closure of the jaws of a pair of Beck dentate aortic clamps An incision just large enough to admit the full caliber of the selected plastic "arterial" catheter is made into the excluded portion of the arterial wall The bevelled tip of the catheter is insinuated between the lips of the diminutive vascular incision, and as the clamp is released, the catheter tip is advanced into the aortic lumen and is passed upward toward the arch Simultaneously the encircling suture is tied down securing hemostasis After triply tying, the long ends of this suture are passed twice about the catheter, are retied, and then again twice about the catheter and retied The terminal end of the catheter is joined to the "arterial" tubing of the bypass system, taking extreme care to eliminate any and all air which might be entrapped A length of polyvinyl tubing may be inserted through a needle puncture made in the wall of the catheter and advanced until its tip lies well beyond the aortic arch for monitoring of the central aortic pressure (Fig 13) A similar technic may be used for insertion of polyvinyl tubing into a venous catheter for monitoring purposes



Fig 12 *A*, Fitch malleable obturators in graduated sizes which aid in the insertion of multifenestrated plastic catheters into vascular lumina *B*, Catheter stiffened by appropriate obturator prior to insertion

Cooling the Patient

Having established the necessary cannulations and having prepared the circulatory bypass system, perfusion is started on a subtotal basis Usually this should be established at about one-third of the estimated full circulatory flow except in the case of small infants in whom, for purely technical reasons, the flow through the perfusion set-up should be relatively greater After establishment of partial circulatory bypass the cooling effect of the heat exchanger in the "arterial" line is brought into play gradually (thus avoiding sudden chilling of the coronary arterial flow) Within 15 minutes in the average case, the central bodily temperature, as measured by an esophageal or preferably, an intrapericardial thermometer, will have been reduced to 68° F (20° C) Usually ventricular fibrillation will have developed by this time, but may be

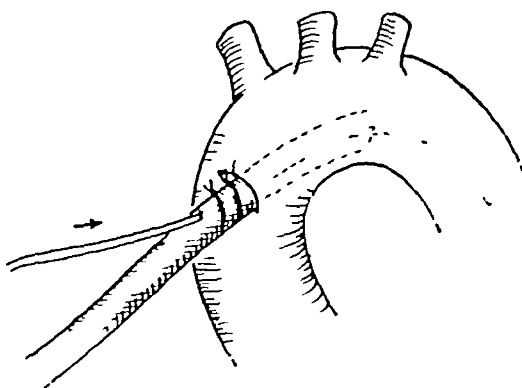


Fig 13 Illustration showing method of inserting the tip of a segment of polyvinyl tubing into the arterial lumen for monitoring of the blood pressure by way of a needle puncture made in the arterial catheter Preferably the catheter should be penetrated before placement, the polyvinyl tubing being advanced subsequently

ignored. The venae cavae are constricted by tightening the encircling tourniquets. The perfusion flow rate may be set at one-quarter of the estimated normothermic bodily flow rate, and the cardiac incision made.

Monitoring the Bypass

A small polyethylene or polyvinyl catheter may be inserted through a puncture wound into any available artery for continuous reading of the systemic arterial blood pressure. Often the most cephalic portion of the exposed aorta offers the most convenient site for such insertion. As mentioned previously the tubing for monitoring of the arterial pressure may be inserted directly through the wall of the catheter which is used for direct return of the oxygenated blood into the ascending aorta (Fig 13) Venous pressures may be measured separately within the two venae cavae by a similar method or perhaps by direct caval cannulation with small diameter polyethylene or polyvinyl tubing inserted through punctures made distal to the site of placement of the encircling tapes.

Blood specimens are taken from one of the caval catheters both before and again after the completion of the bypass for studies of the condition of the particulate blood elements as well as for alterations in the chemical properties of the liquid portion of this fluid tissue. Continuous electrocardiographic and electroencephalographic tracings are visualized on an oscilloscopic screen throughout the operative procedure One must not overlook pertinent physical signs during the bypass. These include pupillary size and reaction to light, the corneal reflex, suffusion of the conjunctival vessels, and spontaneous movements of the extremities. *Oximetry* should be continuously recorded

"Brock" Procedure by Open Technic

When the tender age of the infant or adequate external evidence of extreme hypoplasia of the right ventricular outflow tract or pulmonary artery is deemed sufficient indication for a nondefinitive procedure a Brock type of operation often is selected. The patient is cooled generally by passage of his circulating blood through the heat exchanger At the preselected temperature, usually 68° F (20 C) the perfusion is reduced or interrupted and the venae cavae are closed off by tightening the encircling tapes.

A longitudinal incision is made into the right ventricular chamber making an effort to stay within the *avascular* portion of its anterior wall. Care is taken to avoid unnecessary section of any large coronary arterial branch. The incision of the ventricular wall should be extended fully to the level of the pulmonary valve unless this would necessitate division of a large coronary vessel The *low pressure suction tip* is inserted into the ventricular cavity to evacuate its blood content and to expose the septum.

Should excessive blood emerge from the septal defect it will be obvious either that an unusual amount of collateral circulation exists or that the aortic valve is incompetent. Slowing or stopping the perfusion for 20 to 30 minutes will reduce excessive bleeding from either source. Clamping the aortic root for a short period will abolish any regurgitant aortic flow and also will cut off the coronary perfusion and, hence, the coronary sinus return. We feel it is best to permit a period of vigorous perfusion for several minutes between contiguous episodes of complete systemic or coronary circulatory interruption

One of a series of graduated urethral sounds may be passed through the pulmonary valve to establish whether there is associated pulmonary valvular stenosis or not. If the valve is stenotic, it is felt that a separate longitudinal incision should be made into the pulmonary artery in order properly to expose the valve and its three (or two) commissures (Fig 14A,B) They then *may* be divided with critical accuracy and to the limit that is consistent with maintenance of competence, paying appropriate attention to the structure and configuration of the individual cusps.

The infundibular obstruction when present will be visualized and plans will be made to create a channel between the true right ventricle and the pulmonary valve which will be not over 25 per cent of the size of a normal one. Such a limited passage will provide adequate impedance to prevent excessive passage of blood into the lung from the communicating left ventricle. Thus, the creation of a very large and possibly dangerous left-to-right shunt will be avoided, while the total pulmonary blood flow will be multiplied several times. It is important that final evaluation of the adequacy of the enlarged outflow passage should be established by digital examination after closure of the cephalic half of the incision in the ventricular wall. Otherwise, what may have appeared to be an adequate

passageway before repair of the heart wall may have become reduced by the ventricular closure to a narrow myocardial crevice

The margins of the ventricular incision are approximated with a row of through-and-through continuous mattress sutures, and a more superficial row of running sutures An effort should be made to express any entrapped air from the right ventricle by filling it with saline solution (if it has not already become filled with blood) just before tightening the last applied sutures

plished by the application of external heat to the body

Definitive Correction of the Tetralogy of Fallot

The patient is cannulated as described, and the central temperature is reduced rapidly to 10 to 20° C by circulatory bypass using the heat exchanger in the *arterial* line The flow rate is reduced and the venae cavae are constricted as in the open performance of a Brock procedure However, in this case because the right ventricular outflow tract



A



B

Fig 14 Critically accurate division of the commissures in pulmonary valvular stenosis requires an open technic using a long arteriotomy incision A, Congenital pulmonary stenosis with a "tricuspid" valve structure (autopsy specimen) B, Complete anatomical opening of this valve was performed in the autopsy room No incompetence would have been created in this case

Rewarming the Patient

Rewarming of the circulating blood may be initiated even before starting the closure of the ventricle This is accomplished simply by circulating warm fluid (45° C) about the flow pipes of that heat exchanger which is incorporated in the *venous* line of the extracorporeal circuit

By the time the temperature of the circulating blood has risen to 77° F (25° C) the heart usually will have resumed normal vigorous contractions If ventricular fibrillation should be present and tend to persist, the rhythm readily may be converted to sinus type (or at least to the status of atrial fibrillation) by the administration of lidocaine intravenously (1 5 mg per Kg.) or by electroshock with practically any of the standard defibrillator equipment now available Bypass (and circulatory rewarming) may be terminated when the central blood temperature has attained a level of 91 4° F (33° C) Thereafter, further warming may be accom-



Fig 15: Conventional method of enlarging the stenotic outflow tract and adjacent portion of the pulmonary trunk by the use of a plastic "gusset" The major drawback to this method of handling is the total pulmonary incompetence which it creates (Lillehei, C W, et al: J. Surg, 33:41, 1957)

will have been found large enough and the pulmonary artery estimated to be at least 25 per cent of normal size. The patient being deemed vigorous enough it will have been de termined to complete the operative correction in a single stage.

The right ventricle is incised with or with out interruption of the perfusion as de scribed for the *Open Brock* procedure. The size of the pulmonary artery and valve are established by sounding (or by passage of the operator's finger in an adult patient) If pulmonary valvular stenosis exists it is at tacked directly through a separate incision made in the pulmonary arterial wall. Great care is taken to avoid the production of pul monary valvular incompetence.

The right ventricular outflow tract and the

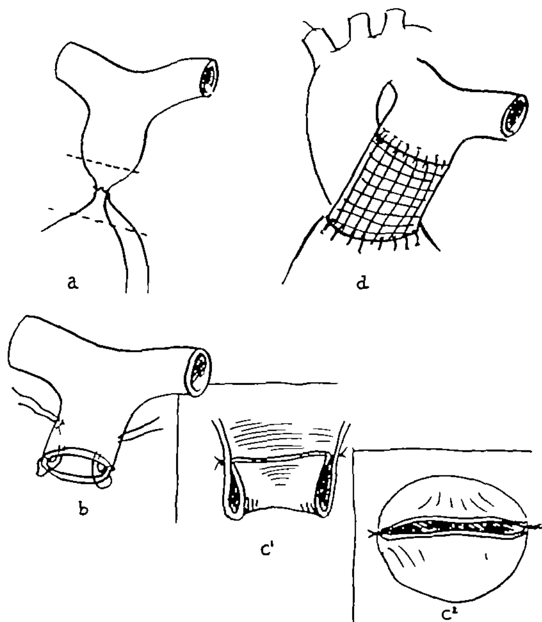


Fig. 16: Recommended (authors') alternative method for enla ging the outflow passage I the region of the pulmonary valve This tech ic provides an efficient "bicuspid" pulmonary valve. a, The entire stenotic region is excised in "sleeve" fashion so that adequate cardiac and vascular lumina become visible Usually there is a dearth of sufficient arterial tissue for direct anastomosis. b, A cuff at the end of the pulmonary arterial trunk is inverted by placement and tightening of two mattress sutures applied through opposite sides of the artery c, This produces an effective bicuspid valve. d Cardiovascular continuity is reestablished by use of an oversized segment of prosthetic tubing. It is important that the prosthetic be rendered impermeable to heparinized blood by preclotting

supraventricular crest are subjected to intense scrutiny An effort is made to establish the largest possible passageway between the true right ventricle and the pulmonary artery However, it is felt that every effort should be made to avoid the necessity for use of a prosthetic patch to enlarge the pulmonary artery at the expense of competence of the pulmonary valve (Fig 15) We feel that it is preferable to use a prosthetic tube to replace the outflow tract and to create a functional bicuspid valve by inversion of the end of the pulmonary artery (Fig 16A,B,C,-D)

Repairing the Septal Defect

In cases of "acyanotic tetralogy" in which it seemingly is permissible to close the usually moderate sized defect completely at one stage, this is done by whichever technic of direct suturing or repair by prosthetic patch seems best to the operator The technical steps, therefore, do not differ significantly from those described in the section Ventricular Septal Defects

In clinically more severe or *blue* cases, we routinely employ a prosthetic foraminal valve applied in such a way as to permit a gradually diminishing right-to-left shunt which will be abolished within a period of from six to twelve weeks These patients nearly always have large septal defects which permit ready fashioning and satisfactory placement of functional valves of *teflon* reinforced *ivalon* sponge Should the defect be unusually small or irregularly shaped, it may be enlarged safely by incising its ventral margin

In the placement of a prosthetic foraminal valve, it is wise to pass the first suture through the plastic ring at the point of maximal valvular opening This is then attached to that point upon the cephalic margin of the defect which lies most directly in the line of ejection of the left ventricle By establishing this point with the first stitch all danger of eccentric placement and possible malfunction of the valve flap will be eliminated It is suggested that the flap be made somewhat shorter than the ring so that it will not impinge or "rub" against a low lying aortic valve cusp The remainder of the circumference of the valve ring may be sutured to the margin of the defect in the fashion usually used for the treatment of ventricular septal defects It is urged that a tangential type of suturing be applied from the left ventricular side, at least in the dorso-caudad quadrant of the circumference of the defect This is known to lie in proximity to the main atrioventricular conduction bundle

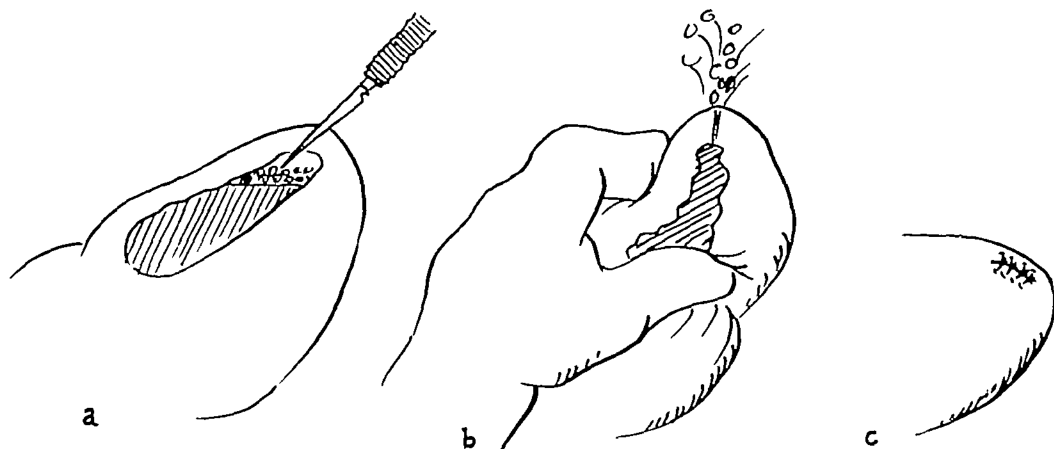


Fig 17 Maximally efficient method for venting of the left ventricle (Nichols technic) a, The apex of the ventricle is elevated and a small stab wound is made into its lumen near the tip b, The lips of this diminutive incision are separated by the blades of a fine hemostat while the operator "squeezes" the ventricles to force any entrapped air (and some blood) out of the opening c, After completely evacuating all air the stab wound is repaired with interrupted sutures

After the plastic foraminal valve has been sutured in place, the surgeon must reassure himself that the valve has been placed so that the flap actually opens into the left ventricle in the line of its outflow and that it tends to close with left ventricular contraction (Fig 10B)

If the artificial valve is truly competent, it is probable at this point that air will have been entrapped within the left ventricular chamber[40]) No danger of arterial air embolism will exist as long as the ventricles are

fibrillating or otherwise nonfunctional However this air must be removed before spontaneous contractions are resumed. We have found it convenient to elevate the cardiac apex so that the entrapped air will ascend into this portion. The air then can be vented efficiently by direct incision of the subapical portion of the left ventricle (Fig 17A,B,C)

The incision of the right ventricle is repaired in thirds beginning with its cephalic portion. The operator explores the capaciousness of the newly constructed right ventricular outflow tract repeatedly as the closure progresses. Should it appear to be constricted unduly the repair may be held in abeyance while further efforts are made to enlarge the outflow channel. *Thinning* of the anterior wall of the right ventricle, or even of the ventricular septum may be deemed advisable (Fig 18A,B C)

The last sutures in the heart wall are drawn tight after any contained air has been evacuated from the right ventricle Rewarming is started by circulating warm water through the heat exchanger which lies in the *venous* line of the extracorporeal circuit. A coordinated rhythm may become established spontaneously or by the intravenous administration of *lidocaine* (usually at about 5 C.) or the ventricles may be massaged or subjected to electroshock. The bypass is maintained at the flow rate which was used during the period of cooling When a temperature of 91 4 F (33 C.) has been reached, the perfusion usually may be terminated and decannulation begun. *Polybrene*[11] is administered by intravenous drip in dosage of 1.5 mg per mg of *heparin* administered (counting the heparin in the blood in the equipment at half value except in the case of infants in whom an effort is made to neutralize only that heparin which actually has been administered to the patient)

Normal clotting time (Lee White) should be obtained within 20 minutes of administration of the polybrene In patients with severe grades of cyanosis defects of the coagulation mechanism are common Here a painstaking and meticulous technic of hemostasis must be instituted before and during closure of the thoracic wound.

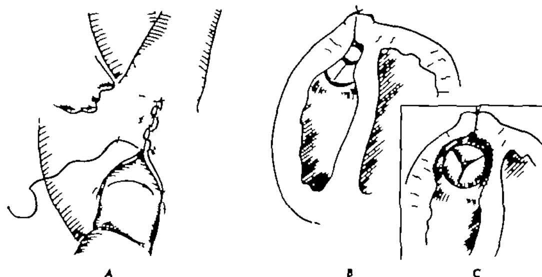


Fig 18 Ensuring the adequacy of the outflow tract A, The upper (cephalad) portion of the right ventricular incision is repaired first Then the operator's index finger is inserted to evaluate the si of the residual passageway B Too small a passageway aft closure of the ventricular wall will necessitate f rther surgery C Often a channel of adequ e siz may be established by "thinning" the adjacent septal wall as well as the right ventricular wall.

Ordinarily the patient becomes "pink" during surgery and remains so postoperatively Nevertheless, if a prosthetic foraminal valve has been used (as a "safety valve" or an auxiliary right ventricular outflow tract) the saturation of the peripheral arterial blood may be found to be less than normal This will serve to confirm the wisdom of maintaining the possibility of a right-to-left shunt in the particular case Eventual closure of the valve abolishes this shunt and renders the correction complete This may be determined readily by follow-up catheterization

after the elapse of several weeks Even though a small to moderate sized pressure gradient may be found to persist between the right ventricle and the pulmonary artery one may take comfort in the knowledge that a moderate degree of pulmonary stenosis is readily tolerable without the necessity for limitation of physical activities

Thus, at one operation (although often only after the elapse of two to three months) most patients with the tetralogy of Fallot may be restored to complete anatomical and clinical health with a relatively low operative risk

References

1 Fallot, A Contribution a l'anatomie pathologique de la maladie bleu (cyanose cardiaque) Marseille med 25 418, 1888

2 Willus, F A and Keys, T E Cardiac Classics C V. Mosby Co, St Louis, 1941, pp 689-839

3 Bennett, L R Sandifort's "Observations" Bull Hist Med, 20 539, 1946

4 Bailey, C P. and Imbriglia, J E In Gould Pathology of the Heart E S Gould, Editor. Charles C Thomas, Springfield, 1959

5 Francia, L Cuore e circ 18 414, 1934

6 D'Autreband, L, Marshall, W R, and Meakins, J C J Clin Invest, 8 123, 1930.

7 Kirklin, J W., et al J. Thoracic Surg 37 22, 1959

8 Leight, L and Davis, L A Am Heart J, 56 129, 1958

9 Barrett, N. R and Dailey, R Brit. M J., 1 699, 1949.

10 Zimmerman, J J Albert Einstein Med Center, 7 77, 1959

11 Blalock, A and Taussig, H B J A M.A 128 189, 1945

12 Potts, W J, Smith, S., and Gibson, S Ibid, 132 627, 1946

13 Ross, R S., Taussig, H B, and Evans, M H Circulation, 18 553, 1958

14 Leeds, S E Am J Surg., 96 234, 1958

15 Sellors, T H Lancet, 1 988, 1948

16 Brock, R C Brit M J, 1 1121, 1948

17 Brock, R C and Campbell, M Ibid., 12 403, 1950

18 Hufnagel, C A Surg Form, Phila 1951, p 246

19 Lin, T K, Diehl, A M, and Kittle, C F Am Heart J, 55 288, 1958

20 Lillehei, C W, et al Ann Surg, 142 418, 1955

21 Kirklin, J W, et al. Discussion Brock, R C, J Thoracic Surg., 37 22, 1959

22 Brock, R C Ann Surg, 136 63, 1952.

22A Brock, R C Am J Card, 3 1, 1959

23 Sealy, W C, Brown, I W, Jr, and Young, W G., Jr Ann Surg., 147 603, 1958

24 Drew, C E, Keen, G, and Benazon, D B Lancet, 1 745, 1959

25 Drew, C E and Anderson, I M Lancet, 1 748, 1959

26 Kay, J H, et al J A.M A., 170 792, 1959

27 Cooley, D A, Belmonte, B A, DeBakey, M E., and Latson, J R Ann Surg, 145 898, 1957

28 Fitch, E A and Denman, F. R Dis Chest, 36 (Dec) 1959

29 Larios, R, Fitch, E A., Blanco, G, and Bailey, C P. Use of an Artificial Foraminal Valve Prosthesis in the Closure of Interatrial and Interventricular Septal Defects 5th Internat Cong Dis Chest Am Coll Chest Phys., Tokyo, Sept 7, 1958 To be published

30 Kolff, W J, et al Cleveland Clin Quart 25 65, 1958

31 Cooley, D A., et al Am Surgeon, 24 870, 1958

32 Bailey, C P, Zimmerman, J, and Likoff, W The Complete Relief of Mitral Stenosis Dis of Chest In press

33 Lillehei, C W J Thoracic Surg 37 272, 1959

34 Benvenuto, R and Lewis, F J Ibid., 37 673, 1959

35 Sirak, H D and Hosier, D M Ibid., 37 1, 1959

36 Taussig, H B J Christian M A India, 32 53, 1957

37. Potts, W J Rev Med Suisse Rom, 76 576, 1956

38 Blanco, G., et al Am. J Card, 2 302, 1958

39. Nunez, L E and Bailey, C P J Thoracic Surg, 37 707, 1959

40 Nichols, H T Venting the Left Ventricle Personal communication to C P. Bailey

41 Weiss, W A., Gilman, J S, Catenacci, A J., and Osterberg, A E. J A M.A., 166 603, 1958

DIVERTICULUM OF VENTRICLE

Charles P Bailey, M D and Dryden P Morse, M D

Definition: True congenital diverticuli of the heart are rare and consist of an outpouching from a ventricle made up of normal appearing ventricular myocardium lined by endocardium and covered by epicardium. They arise apparently nearly invariably from the vicinity of the apex of the heart. They should be distinguished from traumatic or degenerative aneurysms of the ventricle which occur more commonly at other sites and are not composed of heart muscle which contracts normally during systole. Diverticuli also must be differentiated from congenital aneurysms of the coronary arteries which also may occur near the apex of the heart and communicate with a ventricle. The latter do not present subcutaneously and usually are accompanied by a systolic murmur They have physiologic implications because of the shunt. Diverticuli may be contained entirely within the pericardial sac or may penetrate it to lie extrapericardially

Congenital Ventricular Diverticulum

Incidence: Congenital ventricular diverticuli are rare. Only sixteen cases were reported during the 116 years following O'Bryan s first described case in 1839[1]

In these sixteen reported cases the diverticulum was found to communicate with the left ventricle in ten. In two the diverticulum arose from both ventricles, and in four of the reports there is no clear-cut description. In every case the diverticulum arose from or near the apex of the heart, and in ten patients it appeared to penetrate or

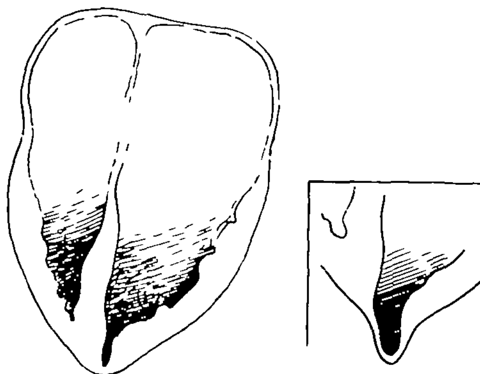


Fig 1 Van d Vijver's concept of progressive blow-out of the ventricular apex producing a diverticulum. (Bailey C. P.: Surgery of the Heart Lea & Febiger Philadelphia, 1955, Ed. 1.)

to displace the diaphragm posteriorly. The sex incidence was equal [2, 3, 4]

Embryology: Embryologists are divided in their etiologic concepts between the theory of Drennan and Van de Vijver[5] which implicates a presumptive increase in left ventricular pressure during intrauterine life as causative of a progressive *blow-out* of the apex (Fig 1) and the theory of Bremer[6] which presents the concept of external traction upon the ventricular apex by an adherent portion of the septum transversum which eventually gives rise to the ventrocephalic portion of the anterior abdominal wall (Fig 2)

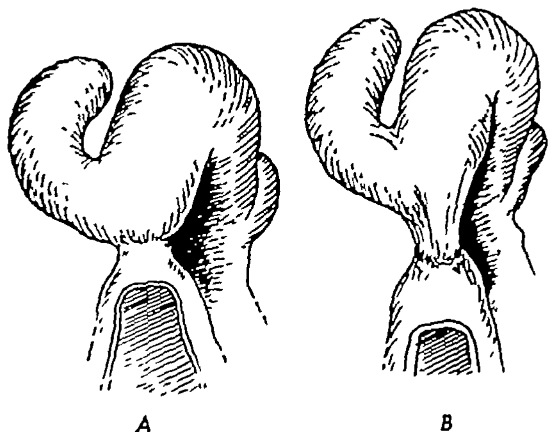


Fig 2 Bremer's concept of mechanism of production of a ventricular aneurysm A, Adherent septum transversum makes traction upon the ventricular apex B, Apex becomes drawn out into a diverticulum (Bailey, C P Surgery of the Heart Lea & Febiger, Philadelphia, 1955, Ed 1)

Clinical Findings: These diverticuli usually present as obliquely disposed pulsating masses which lie superficially under the skin of the epigastrium (in the anterior abdominal wall) In four reported instances they extended down to the umbilicus actually presenting within the umbilical cord They protrude sufficiently in most instances so that they may be grasped partially between the thumb and fingers These protrusions become more prominent on straining and in the erect position, and contract synchronously with the pulsation of the ventricles The protrusion usually is directed from the xiphoid downward and obliquely toward the right (Fig 3)

Physiologic Studies: As mentioned

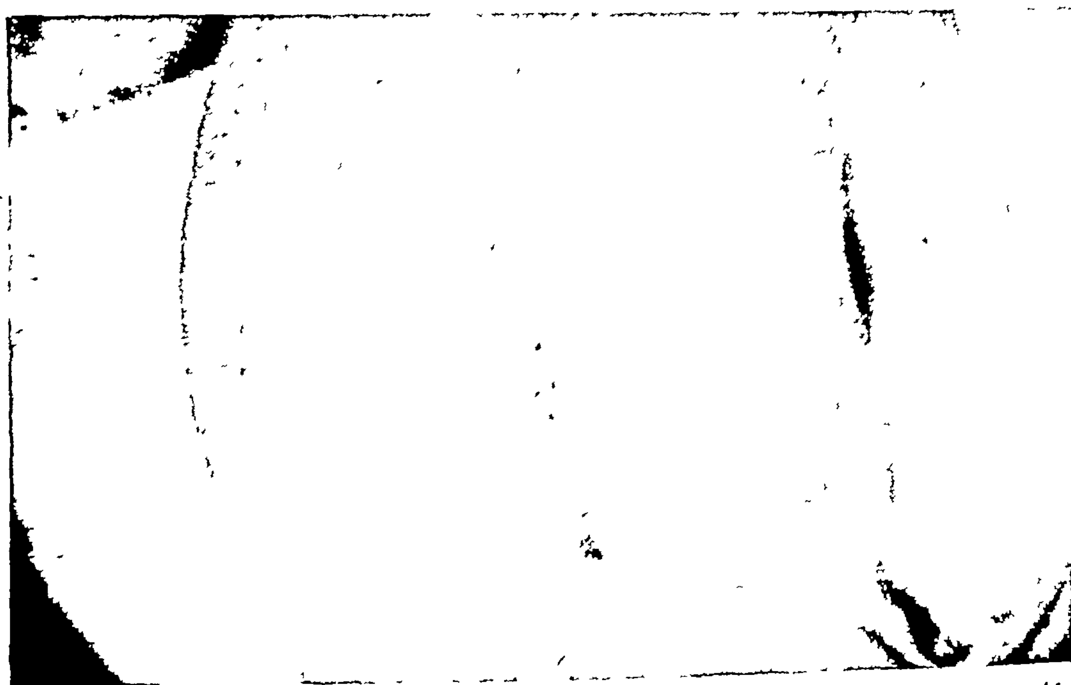


Fig 3 Two week old patient showing pulsating mass in the epigastrium Note defective skin cleavage line running to umbilicus (Bailey, C P Surgery of the Heart Lea & Febiger, Philadelphia, 1955, Ed 1)

these muscular outpouchings usually arise from the left ventricle, but occasionally from the right, or even from both. The latter rare lesion permits a shunt of blood at this level. Other than *in this exceptional situation there is no* physiologic abnormality associated with

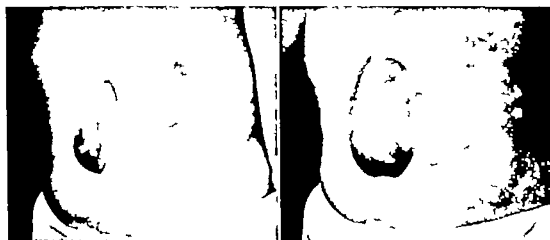


Fig 4 Potts' patient showing larger epigastric pulsating mass. (Potts, W J Surgery C. V Mosby Co., St Louis.)



Fig. 5 Opacification of mass in Skapinker's case by direct injection of contrast medium. A, Lateral roentgenogram. B Posteroanterior roentgenogram during first portion of the i jection. C Posteroanterior opacification of the diverticulum showing direct comm nicatio with the left ventricle (Skapinker et al.; A.M.A. Arch. of Surg., 63:629 1951)

a ventricular diverticulum since the muscular walls are continuous with and contract simultaneously with the ventricles. The nine year old patient reported by Potts[3] presented only nervousness and self-consciousness (Fig 4)

Medical Prognosis: Skapinker[4] reports that the ten patients in his series (from the reviewed literature) who were treated conservatively all died in early infancy except two who died from *rupture of the diverticulum at four* years of age.

X ray Diagnosis: The conventional posteroanterior view may show nothing abnormal Intravenous angiocardiography often is of little value because of poor concentration of the dye within the left ventricle and the diverticulum Direct injection of dye into the pulsat

ing mass, however, may show a communication with the main ventricular chamber as in the cases of Skapinker,[4] Potts[3] and Bailey[2] (Figs. 5 and 6)

Surgical Treatment: The first operated case was that of Wieting[8] who in 1912 replaced a diverticulum within the pericardial sac without an attempt at

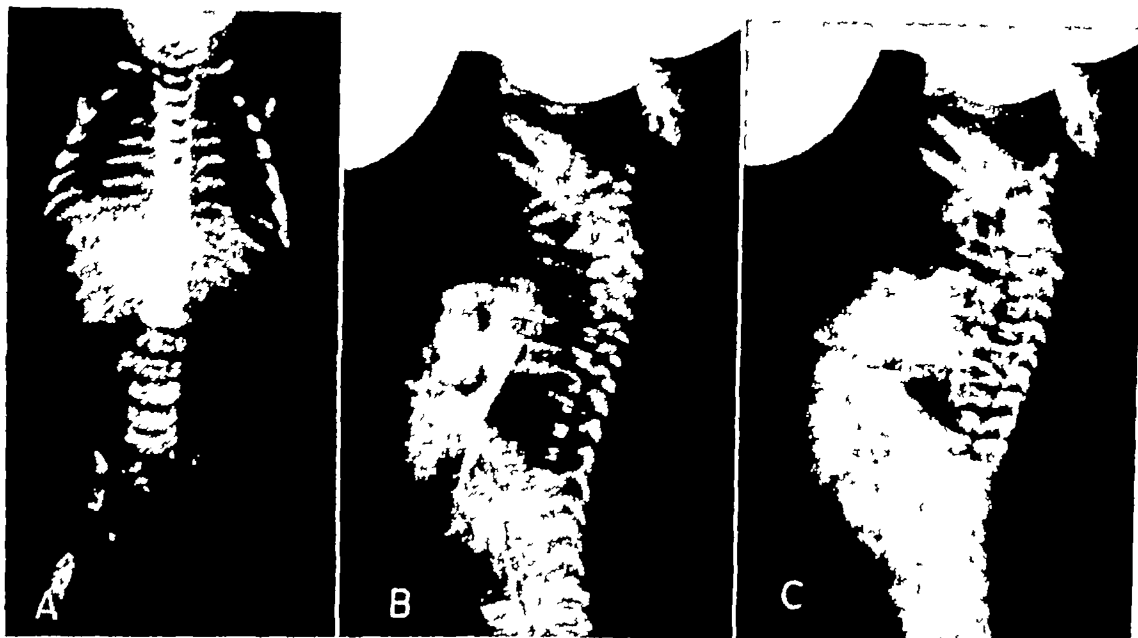


Fig 6 Angiocardiographic demonstration of communication of left ventricle with mass in author's case K W A, Posteroanterior roentgenogram prior to injection B, Lateral roentgenogram obtained early after injection of contrast medium Note filling of right ventricle C, Later serial roentgenogram showing opacification of left ventricle and mass (Bailey, C P Surgery of the Heart Lea & Febiger, Philadelphia, 1955, Ed 1)



Fig 7 Photograph showing division of the diverticulum in author's case K W, distal to a dentate clamp applied close to the ventricular junction After excision the cut end of ventricular myocardium was closed with one row of mattress and one more superficial row of running sutures (Bailey, C. P: Surgery of the Heart Lea & Febiger, Philadelphia, 1955, Ed 1)

removal Roessler in 1944,[9] Skapinker in 1950,[4] Potts in 1952,[3] Bailey in 1954,[2] Gross,[10] Fell,[11] and others more recently have accomplished actual resection All of these patients have survived surgery Early diagnosis and early surgical excision provide a very favorable outlook in cases of congenital ventricular diverticulum

Technic: An incision is made over the mass and the underlying pericardial sac (if any) is dissected and opened (Fig 7) The juncture of the diverticulum with the ventricle is identified and the base of the abnormal structure is cross-clamped with a Morse[7] or other type of myocardial clamp at this site The pouch is amputated and the cut end is oversewn with two layers of continuous fine nonabsorbable sutures After secure hemostasis has been established, the wound is closed in layers.

References

1. Peacock, T B: On Malformations of the Human Heart. J Churchill & Sons, London, 1866, Ed. 2
2. Bailey C. P., Surgery of the Heart. Lea & Febiger Philadelphia, 1955
3. Potts, W J DeBoer A., and Johnson, F R. Surgery 33 301 1953
4. Slapinker S A.M.A Arch Surg 63 629 1951
5. Drennan, M R. and Van de Vijver G T J.M.A South Africa, 2 58, 1928
6. Bremer J L., Arch. Path., 34 1016, 1942.
7. Bailey C. P., Bolton, H E., Nichols, H., and Gilman, R. A., J Thoracic Surg., 35 37 1958.
8. Wieting· Eine Operative Behandelte Hergmissbildung, Deutsche Ztschr F Chir., 114 293 1912
9. Roesler W Erfolgreiche Operative Ent fernung ines ektopischen Hirzdivertikels an einem Neugeborenen, Deutsche Ztschr F Chir., 258 561 1944
10. Gross, R. E. Personal Communication.
11. Fell, E. A J Thoracic Surg., 35 64 1958

SURGERY OF INTERATRIAL SEPTAL DEFECTS

HOUCK E. BOLTON, M.D., AND DANIEL F. DOWNING, M.D.

The subject of congenital defects of the interatrial septum has received extensive attention in recent medical literature. This is occasioned by the facts that the anomaly has come into the realm of curative surgery and is one of the most common of the congenital cardiac abnormalities.

In 1947 Cohn[1] reported the first results of experimental closure of interatrial defects. In 1948 Murray[2] attempted closure in a patient but the result subsequently was proved unsatisfactory by catheterization. The first successful operation to be reported was that of Bailey.[3] Since then, numerous reports have appeared describing various methods of operating on this defect. Outstanding contributions are those of Sondergaard,[4] Bjork and Crafoord.[5] All of these technics are termed *closed* and depend upon tactile sense. Gross[11] describes a method in which he used an *atrial well*. This is an *open* approach, but still depends primarily upon touch, for it, too, is a *blind* procedure. Of all, the technic of atrioseptopexy described by Bailey[3] has probably been most frequently employed, under a variety of terms, and has proved extremely satisfactory.

Repair of atrial septal defects under direct vision was first carried out utilizing hypothermia. Lewis[6] and Swan[7] pioneered this phase of the development of surgery.

Gibbon[8] made the first report on the use of extra-corporeal circulation and direct vision for the successful closure of an interatrial septal defect. Subsequently numerous papers relating to similar treatment with pump-oxygenators have been published.

Abnormality of development of the interatrial septum may result in various types of defects which will allow a communication between the two atria. The commonest type is the "ostium secundum" defect. One may assume that in this condition either the septum primum has undergone excessive resorption or the development of the septum secundum has failed. In either event, there results an aperture which may be considered a greatly enlarged foramen ovale. There is no foraminal valve to prevent the trans-septal passage of blood.

The "ostium primum" defect is considered to be caused by failure of the septum primum to fuse with the endocardial cushions (primordia of the atrioventricular valves).[9,10] Watkins and Gross[11] consider these defects as "endocardial cushion defects" and group them with other developmental abnormalities which occur in this area such as atrioventricularis communis. Under such circumstances, the atrioventricular valves are prone to be deformed and consequently often incompetent. In defects of this type, often there is complete absence of tissue which would represent the interatrial septum (Fig. 2). In some cases there are simply lace-like strands of septal tissue traversing the defect.

The defect of the septum secundum type may be located variously (Figs. 1 and 3). In approximately 50 per cent of the patients there will be a defect which is either centrally located or else high in the septum near the mouth of the superior vena cava.[12] In approximately 10 per cent of the patients with interatrial septal defects, there is associated anomalous pulmonary venous

drainage in which the pulmonary veins from the right lung drain into the right atrium This anomalous drainage is usually associated with a defect located posteriorly in proximity to the venous ostia

right atrium into the left would ensue Elevated atrial pressure will tend to produce dilatation of the atrial chamber and increase the size of the opening

If there is no complicating element in the presence of an atrial septal defect,



Fig 1 Septum secundum type of interatrial septal defect with residual rim of septal tissue

Pathophysiology

Left atrial pressure normally is higher than that of the right atrium The foramen ovale, when this is patent, is so designed that the foraminal valve will prevent blood flow from the left to right atrium under ordinary conditions If the pressure in the right atrium should for any reason exceed that in the left atrium, then a flow of blood from the

the pressure in the left atrium is normal and there is allowed a shunt of blood from left to right This results in a dilatation of the right atrium and an increase in pulmonary flow followed by hypertrophy of the right ventricular wall as well as dilatation of the chamber Because of increase in the flow of blood through the pulmonary vascular bed, by means of a mechanism at present incom-

pletely understood, there results in some patients pulmonary hypertension. As the pulmonary resistance increases, the left to right shunt becomes decreased and there is an increase in the systemic flow. This sets up a cycle of events wherein the right ventricle fails to empty properly. The right atrial



Fig. 2: Septum primum type of defect with only a lace-like remnant of the septum being visible.

pressure gradually increases and a balanced shunt between the two atria, or finally a right to left shunt develops. Cyanosis appears because of this right to left shunt.

The Clinical Picture

The diagnosis is established much more frequently in older individuals than in infants or young children, leading to the view that, in the majority the defect probably is silent early in life.

The most common symptom is easy fatigue and, eventually nearly all patients will have this complaint. Shortness of breath is almost as prominent. The impression is widespread that respiratory infections are a feature of the natural history. In our group of patients this has not been true.

The physical signs are not striking. The majority of patients are normally developed. A gracile habitus is no more frequent than in any other significant defect. The sternum and left anterior thorax bulge in many patients. A thrill is found in a relatively small number. The rhythm is almost invariably normal. The second sound at the base, to

the left of the sternum, is commonly accentuated and split. A systolic murmur is present in all but a few individuals It may be heard best at any level from the second to the fourth left interspace, and usually is softer than the murmur of ventricular septal defect. It may be loud and harsh An early or early-to-mid diastolic murmur frequently is heard at the apex or along the left sternal border. Cyanosis is present

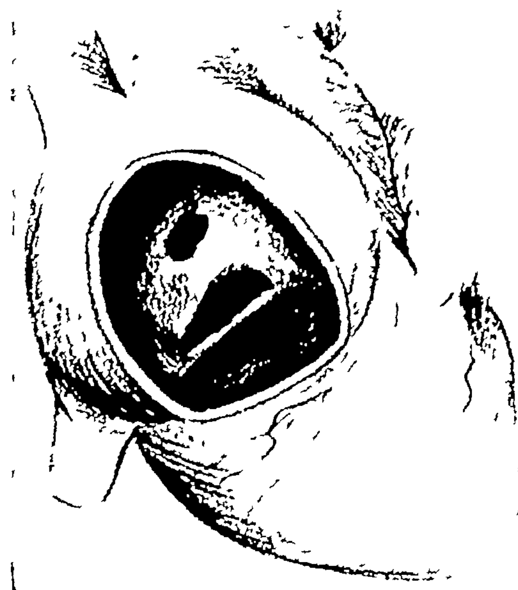


Fig 3 Small interatrial defect of septum primum and secundum types coexisting

only in those patients with marked pulmonary resistance and reversed shunts

Electrocardiography offers help in diagnosis In 70 per cent of the patients there is an rsR[1] pattern in the right precordial leads

The pulmonary vascular markings are accentuated on roentgenograms The main pulmonary artery and its right and left branches are dilated. Marked dilatation of the branches is almost diagnostic of atrial septal defect The heart may or may not be enlarged. Usually, in the right anterior oblique view there is anterior prominence of the cardiac shadow.

The value and necessity of cardiac catheterization cannot be overemphasized in the case of suspected interatrial septal defects As we have previously stated,[12] as much detailed information as is possible should be obtained prior to surgery. We are in disagreement with Blount, *et al.*,[13] who hold that, in general, cardiac catheterization is not necessary A clinical diagnosis which might be erroneous or incomplete in only one per cent of operated patients is not acceptable when a simple, safe means of establishing a diagnosis is available.

Surgical Treatment

Selection of Patients: Since the surgical correction of interatrial defects was first performed in 1952, many changes in concept have materialized as broader experience has been gained This experience has resulted in a greater discrimination in choice of patients for operation

The ideal patient is one who is two years of age or older, with a septum secundum defect allowing a left to right shunt of 1 5 liters of blood per minute or more The pulmonary arterial pressure is normal or, if elevated, is considerably lower than systemic. Surgery performed upon this class of patient will be attended by an extremely low operative mortality and an excellent clinical result Associated anomalous pulmonary veins and/or mitral stenosis are not deterrents to surgery nor do they alter the prognosis

The problem patients are those few patients who have a septum primum type of defect regardless of the pulmonary vascular resistance, and those patients with an ostium secundum defect and marked pulmonary hypertension. This latter condition will sooner or later produce a right to left shunt and cyanosis

There is some hope, however, for

patients with a balanced or right to left shunt. Our use of an "artifical foramen ovale" made of *nylon sponge* (Interventricular Septal Defects, Figure 12 page 46) has encouraging features and may ultimately provide better operative results in these poor risk patients. Cases of atrioventricularis communis must also fall into the group of problem cases.

Technic: Each of the three methods most commonly used for the surgical correction of interatrial septal defects has individual merit. These are the *closed* or atrioseptopexy method designed by Bailey the open or direct method using hypothermia, and the open technic using extracorporeal circulation All three, when applied to the ideal patient have produced excellent results. Probably the lowest operative risk is experienced with atrioseptopexy followed by the hypothermic technic and that with extracorporeal circulation

Each has its advantages and the technic should be selected according to the need of the particular patient.

The closed or tactile method consists in the placement of sutures in such a way that the defect is obliterated without opening the heart. Sondergaard described a method in which a single suture is placed circumferentially about the defect when tightened the communication is closed. This technic is used infrequently in the United States, but commonly in Europe.

The method described by Bailey as *atrioseptopexy* consists of suturing the redundant right atrial wall to the edge of the defect. In selecting this method it must be borne in mind that a very thorough knowledge of atrial anatomy and orientation by tactile methods must be had. In experienced hands it has proved the safest and most efficient method for the treatment of the "ideal patient" for surgery Atrioseptopexy has the distinct advantage of requiring no special equipment or large quantities of blood.

The operation is performed with the patient in the supine position. After induction of anesthesia and insertion of the endotracheal tube, a right inframammary incision is carried through the fourth intercostal space. The internal mammary vessels are individually ligated and divided and the fourth costal cartilage is divided The lung is retracted laterally and the pulmonary veins are inspected for anomaly

The pericardium is opened anterior to the right phrenic nerve. The interatrial groove is dissected out and a nylon purse string suture is placed widely around this area followed by the insertion of a mattress suture across the dissection point. A stab incision is then made into the left atrium and the ungloved left index finger is inserted for exploration of both atria, the mitral and tricuspid valves, and the septal defect. If a septum secundum defect alone is present its closure is then effected.

The tip of the index finger engages the edge of the defect and the thumb presses the outer wall of the right atrium so the two structures come together The suture passes through the atrial wall, the rim of the defect and out through the right atrial wall Similar sutures are placed around the entire circumference of the defect. When these sutures are tied, the defect is closed (Fig 4A D) The index finger is removed from the interatrial groove and the closure further secured by a row of continuous everting sutures using 000 cotton.

During the placement of the sutures in the edge of the defect, care is exerted to avoid placement of a suture which may obstruct the flow of blood from either vena cava into the right atrium. The pericardium and right hemithorax are flushed with normal saline solution to remove any blood clots. The entire operative area is inspected to insure hemostasis and the pericardium is closed loosely A drainage catheter is placed in the right pleural space and after inflation of the right lung, the chest is closed in a routine fashion.

We have not found it necessary to heparinize these patients postoperatively

Mitral stenosis may be corrected prior to closure of the defect when the lesions coexist We have treated eight such cases successfully It does not add

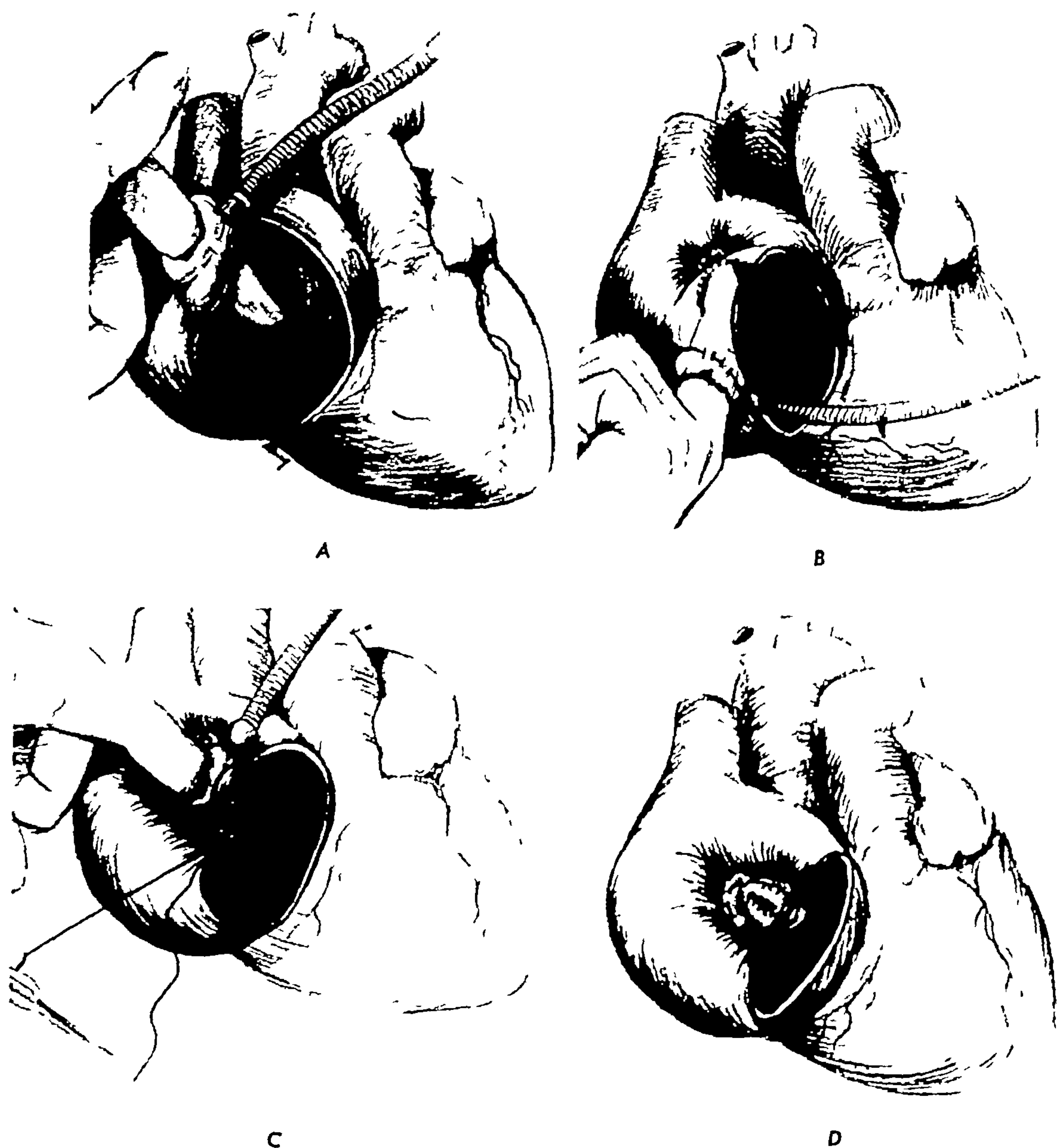


Fig 4 Atrioseptopexy A, Exploration of atrial septal defect and all other structures of both atria, tricuspid, and mitral valves, and pulmonary veins prior to atrioseptopexy B, First suture in place bringing right atrial wall to edge of atrial defect C, Progressive sutures bringing atrial wall to edge of atrial septal defect D, Completion of sutures around edge of defect with "cut-out" showing interior of right atrium and closure of site where the index finger was located

to the operative risk of the repair of an atrial septal defect of the secundum type

Associated anomalous pulmonary veins which empty into the r' m

can be corrected by using the technic of atrioseptopexy and simply suturing the atrial wall to the edge of the defect in a semicircular fashion so that the veins are excluded from the right atrium and shunted through the defect into trium (Fig. 5). We have succ ormed sixteen such proc

Results experienced by the use of atrioseptopexy have been most gratifying. The operative survival in the first 107 patients classified as the ideal patient has been 96.3 per cent (four deaths). Total abolition of the shunt has been accomplished in 95.0 per cent of the cases.[12]

The problem cases cannot be satisfactorily operated upon by atrioseptopexy. Extracorporeal circulation is used in such instances.

occasional septum primum defect encountered which was not diagnosed preoperatively, this lesion cannot be repaired properly by using the closed or tactile methods.

The disadvantages to the use of hypothermia are few. It requires a longer operative time because of the induction of and recovery from hypothermia. The safe duration for open surgery is considered generally to be ten minutes or less as compared to the almost un-

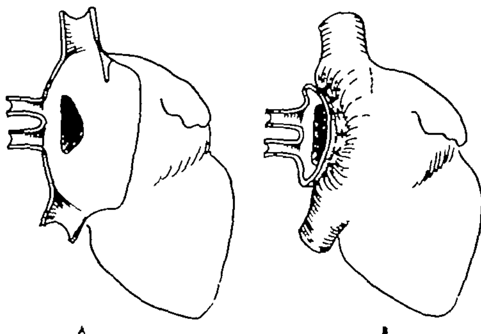


Fig. 5. Atrioseptopexy: A, Anomalous drainage of right pulmonary veins into right atrium. B, Semicircular row of sutures bringing atrial wall to edge of the defect, shunting right pulmonary venous flow through the defect into the left atrium.

Hypothermia: There are numerous advantages in using hypothermia. Its simplicity in the amount of equipment required as compared to extracorporeal circulation is very important. It does not imply the tactile skill and experience required in atrioseptopexy in order to obtain proper closure. It does not require the very large amount of freshly drawn blood which is necessary for performing the operation with extracorporeal circulation. It is sometimes possible by this means to repair the

limited time permitted by the use of extracorporeal circulation. There is always some danger of coronary or cerebral air embolization when open technics are used. Of course, this is precluded by the use of closed methods.

Induction of hypothermia may be accomplished by immersion of the patient in ice water, placing the patient between two blankets containing coils through which an alcohol solution at 0° C. (or slightly below) is circulated and a third, and quite rapid method,

which consists of an extracorporeal circulation of blood from a vein through a coil submerged in ice water and back to the venous circulation of the patient. A number of modifications or combinations of these technics have been used.

The blanket method is the slowest in reducing the temperature to the desirable level, requiring about one to three hours. Immersion requires about thirty minutes to an hour The extracorporeal cooling of the blood requires even less time In each case the rectal temperature is recorded and when it reaches 86 to 87 8° F (30 to 31° C), induction is stopped The temperature will continue to fall or "drift" to 78 to 82 4° F. (26 to 28° C). It is generally felt that if the temperature is allowed to drop below 77° F (25° C), there is an increased hazard.

Technic *The open operation with hypothermia* is begun with the induction of anesthesia and endotracheal intubation Electrocardiographic leads are connected to a cardioscope so that continuous monitoring of the cardiac rate and rhythm is possible Blood pressure is recorded by cannulating an artery and direct manometric readings are continuously observed Body temperature recordings are usually rectal Hypothermia is then induced The surgery is begun when it is felt that by the time the heart is exposed, the appropriate temperature will be reached

The chest may be entered by a transternal bilateral thoracotomy incision at the fourth intercostal space or a vertical sternal splitting entrance into the mediastinum for pericardiotomy without opening the pleural spaces We prefer the latter approach, avoiding the pleurotomy This makes for a more benign postoperative course The pericardium is opened widely, a purse string is placed about the right atrial appendage, incision is made into the right atrium and an exploring finger is inserted to obtain as clear a picture of the nature of the existing defect as possible

An umbilical tape is passed around the superior and inferior venae cavae to occlude the flow of blood into the heart when necessary If one uses prostigmin to help prevent ventricular fibrillation, the root of the aorta is clamped and 0 25 mg of *prostigmin* is injected proximally This will perfuse the coronary arteries in about five seconds Then the aortic clamp is released

The tapes around the cavae are tightened and a clamp is placed across both the aorta and pulmonary artery A generous vertical incision is made in the right atrial wall Blood is aspirated to the level of the septal defect and the atrial chamber is inspected If blood is allowed to remain in the left atrium, the possibility of air entering the left ventricle is lessened

The septum secundum type of defect is then closed with a continuous suture using as *fine gauge nylon* as the delicacy of the septal rim tissue will allow Just before the final two sutures are placed, saline is allowed to flow into the left atrium to displace any air and then the closure is completed

If the defect is so large as to preclude bringing the edges together or if a septum primum defect is present, a thin patch of compressed *ivalon sponge* is sutured into the position that a normal septum should assume

The right atrial wall is elevated by previously placed traction sutures, the atrium is filled with saline solution and a clamp is used to approximate the edges of the atrial incision

The two caval tapes are removed and the apex of the heart is elevated so any air in the left ventricle will be at the apex A #17 gauge needle is used to aspirate the apex of the ventricle to ensure the removal of any existing air This maneuver should be performed as rapidly as possible Following this the clamp is removed from the aorta and the pulmonary artery

The atrial incision is closed with a continuous everting mattress suture and a second row of simple over and over sutures The atrial clamp is then removed

The heart usually resumes a normal rhythm However, if ventricular fibrillation develops, electric shock and massage will usually convert the heart to a normal rhythm We have found the use of *lidocaine* (1 0 mg per kilogram body weight) helpful in converting a persistent ventricular fibrillation It may be injected directly into the left ventricle

The rewarming may begin as soon as the right atrium is closed, This may be accomplished by diathermy, coil blanket, or heating the extracorporeal coiled tube connected into

the venous circuit. It will require thirty minutes to one or more hours to bring the temperature back to normal, depending on the size of the patient and the rewarming method used.

Hypothermia has been selected as the method of choice by Swan and others for the ideal patient. The surgical risk using this technic has been 5 6 per cent in Blount and Swan's[13] series as compared to 3 7 per cent with atrioseptopexy in our experience. It is an effectual method of repair and may be the method of choice for the less experienced operator This method should not be used for the problem case because the amount of time which the heart may be kept open for definitive surgery is limited to about ten minutes. Often it is necessary to have more time for the closure of the defect in the problem case and accordingly extracorporeal circulation must be used.

Extracorporeal Circulation: In the future the apparatus for extracorporeal circulation will undoubtedly be perfected to a point that its use will not add appreciable risk to any surgical procedure. Until such time we are using extracorporeal circulation preferentially in the problem cases as defined. These have septal defects which cannot be closed by atrioseptopexy and the limited time allowed for open heart surgery with hypothermia is inadequate.

The surgery for repair of an atrial septal defect using extracorporeal circulation is performed through a sternal splitting incision as described with hypothermia As soon as the heart is exposed, the patient is heparinized. Both the superior and inferior venae cavae are cannulated for the removal of venous blood to pass through the oxygenator A femoral artery or the ascending aorta is cannulated to receive oxygenated blood. When the by pass connections are complete, umbilical tapes around the venae cavae are tightened to prevent venous blood from entering the heart. After a few beats the heart will empty its contents into the aorta. A longitudinal incision is made into the right atrium. The chamber and the defect are inspected Since in the septum primum defect there may be a cleft in the septal leaflet of the mitral valve, it is sought for and sutured if present. In these cases there is little or no septal tissue present so that a thin sheet of compressed *ivalon sponge* is cut to approximate size and sutured into the position which the interatrial septum normally occupies. A continuous fine nylon suture is used The cardioscope or electrocardiogram is watched to be sure a suture is not passed through the conduction tissue. In order to prevent air embolism a sizable (1 to 1.5 cm.) incision is made in the contra dependent portion of the left ventricular wall. This is a most important step, for air trapped in the left side of the heart may result in fatal cerebral or coronary arterial embolism. The ventricular wound is sutured finally

Following closure of the defect the right heart is filled with saline solution and a clamp applied to close the right atrial incision The incision is closed with a row of continuous everting mattress sutures and a second row of continuous simple sutures. The tapes around the vena cavae are removed and the flow through the pump-oxygenator is slowly reduced as the heart begins to resume its function. When the blood pressure is near normal the extracorporeal circulation is stopped, the anesthesiologist resumes control of respiration. The heart function and the electrocardiogram are observed for a short period to be certain that there has been no injury to the conduction mechanism. *Polybrene* or *protamine* is given intravenously to counteract the effect of heparin and the caval and arterial cannulae are removed Meticulous care must be given to hemostasis at the time the chest is entered and prior to closure in patients who are heparinized for extracorporeal circulation. A two way catheter is placed in the pericardium prior to closure of the chest so that the sac may be drained and also irrigated with normal saline solution to prevent accumulation of blood and resultant tamponade

The results obtained in the cardiac surgical centers have been variable in this group of problem cases. Cooley[14] has had very good results in a series of twenty four patients with an ostium primum type of defect with only three

deaths (12 per cent). Likewise he has had excellent results in closing the ostium secundum defect utilizing extracorporeal circulation He has a series of 119 cases with an operative mortality of only 4 per cent Out of this group there were 100 patients less than twenty five years of age with no pulmonary hypertension in whom there were no deaths.

Conclusions

The *ideal patient* with an atrial septal defect may be treated surgically in one of three ways These are the closed technic using atrioseptopexy, open heart surgery utilizing hypothermia, and open surgery using extracorporeal circulation.

The results of atrioseptopexy are very good and present slightly less risk surgically. The logistics of this technic present less of a problem than the open technics (Table 1).

TABLE 1

Ostium Secundum Defect Surgical Results

	Number of Cases	Operative Deaths
Atrioseptopexy	107	4 (3 7%)
Hypothermia		
Blount, et al[11]	125	7 (5 6%)
Pump Oxygenator		
(1) Combined data reported by Kieffer[15]	99	11 (11%)
(2) Cooley[14]	119	5 (4 0%)

The hypothermic technic applied to the ideal patient presents fewer problems in terms of the amount of blood required for the procedure and less equipment than extracorporeal circulation

Extracorporeal circulation may be utilized for the ideal patient. The risk of surgery is rapidly becoming less in this procedure because the technic of the extracorporeal circulation is rapidly becoming more refined as a result of the correction of numerous faults which existed in the early experience with pump oxygenators It permits direct vision and sufficient time for repair of any defect encountered at surgery which may not have been recognized preoperatively

The cases described here as the "problem cases" (Table 2) can be treated surgically only by means of the pump-oxygenator technic for they cannot be managed technically by atrioseptopexy, and there is inadequate time for their repair when hypothermia is used At best the operative risk in these patients is still rather high

TABLE 2

Ostium Primum Defect Surgical Results

	Number of Cases	Operative Deaths
Hypothermia		
Combined series reported by Kieffer[15]	14	9 (64%)
Pump Oxygenator		
(1) Combined series reported by Kieffer[15]	20	9 (45%)
(2) Cooley[14]	24	3 (12%)

It is quite probable that in the future the difficulties with extracorporeal circulation may be overcome to the point that all types of interatrial septal defects will be closed by using a pump-oxygenator. It is the only method which provides an opportunity of treating both the "ideal patient" and the "problem case."

References

1. Cohn, R Am Heart J., 33 453, 1947

2 Murray, G. Ann Surg, 128 843, 1948

3 Bailey C. P et al: Ann. Int. Med 37 888, 1952
4 Sondergaard, T., Gottsche, H., Ottosen, P., and Schultz, U J Acta chir scandinav 109·188, 1955
5 Bjork, V O and Crafoord, C. J Thoracic Surg., 26 300, 1953
6. Lewis, R. J and Taufic, M. Surgery 33: 52, 1953
7 Swan, H., Zeavin, I Blount, S. G., Jr., and Virtue, R. W.. J.A.M.A., 153 1081 1953
8. Gibbon, J H.. Minnesota Med., 37 171, 1954
9 Patten, B M.. Am. J Path 14 135 1938.
10 Edwards, J E.. Pathology of the Heart Gould, S E., Editor Charles C. Thomas, Springfield, 1953
11 Watkins, E., Jr and Gross, R. J Thoracic Surg., 30·469 1955
12. Bolton, H., Lazarides, D., Goldberg H., and Downing D F Arch. Surg., 74 351 1957
13 Blount, S. G., et al. J.A M.A., 169 210, 1959
14 Cooley D A Personal Communication.
15 Kieffer S. A. Dis. Chest, 34 424 1958

SURGERY FOR AORTIC STENOSIS

CHARLES P BAILEY, M.D., AND DRYDEN P MORSE, M.D

Of the cardiac valvular lesions aortic stenosis is second only to mitral stenosis in the frequency of its clinical and pathologic occurrence. Commonly it is caused by rheumatic fever and is found pathologically in at least one-quarter of all cases of rheumatic heart disease. It also may result from degenerative calcific changes in the valve, and is occasionally of congenital origin. Aortic stenosis due to rheumatic disease usually becomes symptomatic in the fifth or sixth decade, and is more common in men than in women by a ratio of two to one. This is in contrast to the predominant incidence of mitral stenosis in females.

The exact method of correction in aortic stenosis is dependent upon the specific one of the various basic pathologic entities which is encountered. This can be determined best after the aorta has been opened for direct vision repair

Pathology of Aortic Stenosis

Congenital: Aortic stenosis of congenital origin may be present as part of a general hypoplasia of this region, or the stenosis may be limited to the leaflets alone In the latter instance it may take the form of a cap-like fusion of all

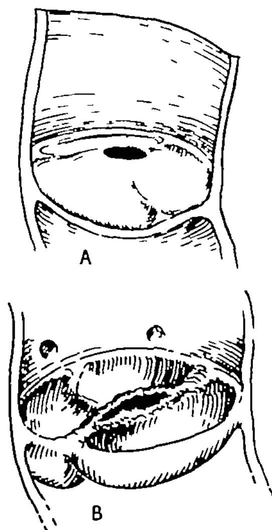




Fig 1 A, Congenital fusion of all three commissures produces a valve which is a thin cone in diastole and a balloon-like dome with a central perforation in systole. B and C, Diagram and photograph of the usual type of congenital aortic stenosis in a bicuspid valve. Both of the main commissures are somewhat fused and there may be a third rudimentary raphe incision of which will surely produce insufficiency (Bailey C. P.: Surgery of the Heart Lea and Febiger Philadelphia 1955.)

three commissures There may be absence of one commissure with bicuspid formation of the valve In many of these cases a rudimentary raphe may be seen to *divide* the expanse of the larger of the two valve cusps (Fig 1A,B,C) mately 1 cm beneath the level of attachment of the valve leaflets

A bicuspid aortic valve may be present without either obstruction or congenital fusion of its two commissures In later life such bicuspid valves



Fig 2 A calcific and stenotic valve Fusion of the commissures is the least part of the extensive pathological process which has converted this valve into a "rockpile" The calcification is predominantly subendothelial and within the concavities of the cusps Some free material, however, is on the channel or ventricular aspect of the valve particularly beneath the commissure at the lower right Calcific masses in this and similar cases may rise high enough to block and hide the coronary ostia and impede their cannulation (Bailey, C P and Likoff, W A M A Arch Int Med 99, 859, 1957)

Poststenotic dilatation of the ascending aorta is common in congenital aortic stenosis This is not true in subvalvular stenosis which most commonly aortic stenosis presents as a relatively thin diaphragm or circumferential ridge lying approxi- may become infiltrated with calcium and then may become stenotic due to stiffening

Degenerative or "Arteriosclerotic": Commonly a history of rheumatic fever is not obtainable in patients

with arteriosclerotic aortic stenosis. Classically in this type of pathology there is no commissural fusion. The circulatory impedance in arteriosclerotic valvular stenosis usually is due to calcification and fibrosis of the valve leaflets with thickening and stiffening

Rheumatic: In rheumatic aortic stenosis there will have been inflammatory involvement by rheumatic endocarditis of the leaflet surfaces This causes, by normally subject. This calcification may occur subendothelially in the substance of the valve leaflets, commonly thickening the central portions of the concave aspects of the leaflet edges or gradually filling in the sinuses of Valsalva. Frequently calcification is more marked in the region of the fused commissures. In the more advanced cases the calcification may involve the entire valve and extend into the aortic wall itself In



Fig. 3 The under surface (ventricular aspect) of a calcific aortic valve (upper center) and of mitral valve (lower center) into the aortic (septal) leaflet of which the calcific process has extended Also note in the upper portion of the picture how the calcific process has extended below the aortic valve into the fibrous portion of the interventricular septum in the area of the conduction bundle. (Bailey et al.: The Surgical Treatment of Aortic Stenosis, Encyclopedia of Thoracic Surgery Courtesy of Springer Verlag)

later organization of the inflammatory exudate, both cross-fusion of the leaflets at the commissures, and thickening and scarring of the valve edges. Both of these processes tend to promote a contractural stricture of the valve opening with a gradual decrease in the size of the orifice. In 90 per cent of these patients *calcification develops presumably* in relation to the hemodynamic stress and trauma to which the aortic valve is

such cases the valve becomes converted essentially into a *rockpile* (Fig 2) The calcification may build up from the surface of the valve as stalagmites which are bare of any endothelial covering The deposition may take place along the wall of the outflow tract in the subvalvular area. In extreme cases it may be continuous with calcification in the mitral valve (Fig 3)

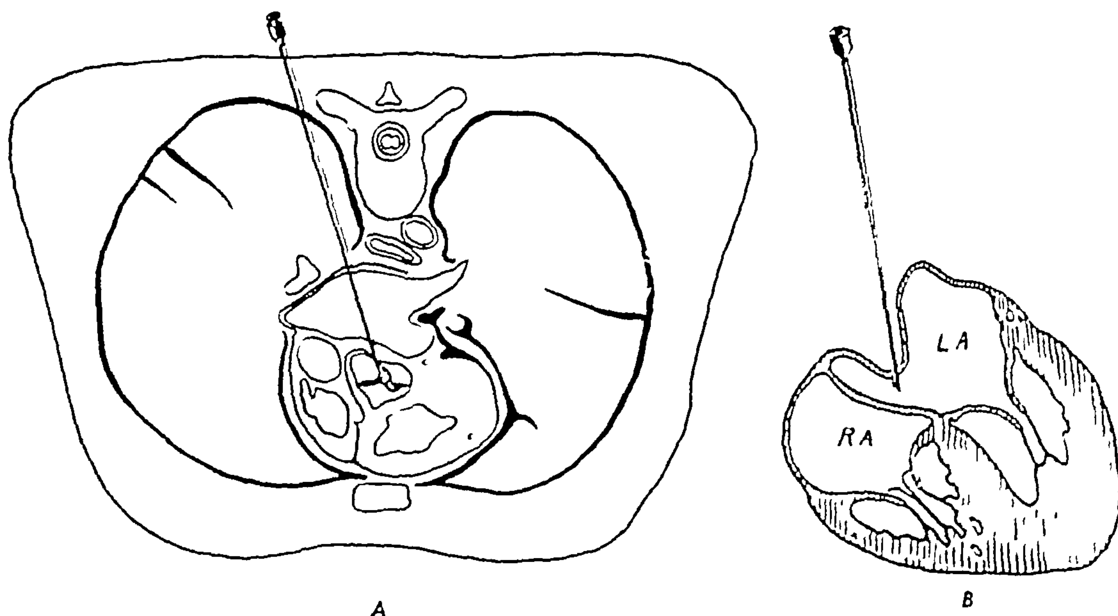
In pure aortic stenosis the left ven

tricle characteristically undergoes a concentric hypertrophy or thickening of the muscular wall without commensurate increase in the size of the left ventricular chamber In early cases there will not seem to be much enlargement of the heart radiographically, although the characteristic "boot" shape may become apparent later (Fig 4A,B)



A B

Fig 4 A and B, The posteroanterior and lateral views of the heart on x-ray show the concentric hypertrophy of the left ventricle with an elevation and rounding of the apex in the posteroanterior view In aortic insufficiency the left border of the dilated heart slants down in a more triangular fashion to a "drooping" apex



A B

Fig 5 A, In the transthoracic method of left heart catheterization described by Bjork and modified by Fisher, a long needle is inserted posteriorly to the *right* of the vertebral column in the patient lying prone on a fluoroscopic table B, A sharp six inch No 18G thin walled Becton and Dickinson needle is inserted into the left atrium As the needle pushes against the left atrial wall, a definite resistance can be felt, followed by a popping sensation on entry

Pathophysiology of Aortic Stenosis

Because aortic stenosis impedes ventricular ejection the heart compensates by increasing (1) the pressure behind the valve, and (2) the systolic time per minute for ejection. It has been demonstrated that untoward hemodynamic effects due to narrowing of the valve do not become apparent until the effective valve opening has been reduced to approximately 25 per cent of its normal size (0 8 cm.) When the valve orifice



C



D

Fig 5 C Photograph of the begi ing of a actual left heart catheterization. The paper emerging from a m bi-channel direct writing record is in the left pper corner of the picture D Afte the needle is placed i the left atri m, a thin polyethylene catheter is inserted through the needle into the left ventricle The three-way stop-cock allows irrigation t preve t clotti g The cardioscope (rea) is mo itored throughout the proced re

becomes less than 10 per cent of its normal size, the patient usually develops impending left ventricular failure, reflected at first, by increased pressure within the left atrium which extends into the pulmonary veins Anginal pain tricle. There is serious reduction in the patient's ability to increase his cardiac output upon demand so that unusual exertion may cause temporary hypotension and syncope.

Direct pressure measurements across

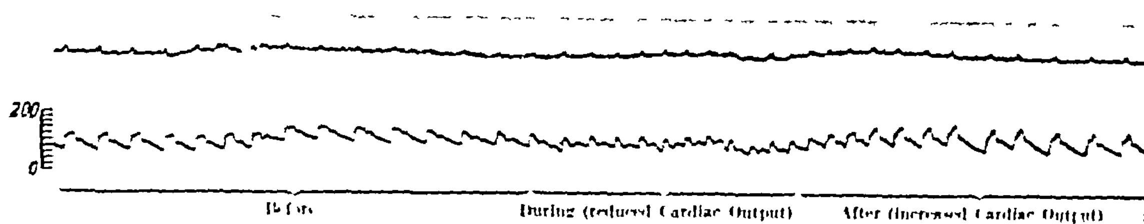


Fig 6A Brachial arterial tracing obtained in a patient with aortic stenosis before, during, and after the Valsalva maneuver *(straining during breath holding)* Note disappearance of anacrotic notch during period of reduced stroke output and exaggeration during subsequent period of corrective overshoot (Bailey, C P Surgery of the Heart, Lea and Febiger, Philadelphia, 1955)

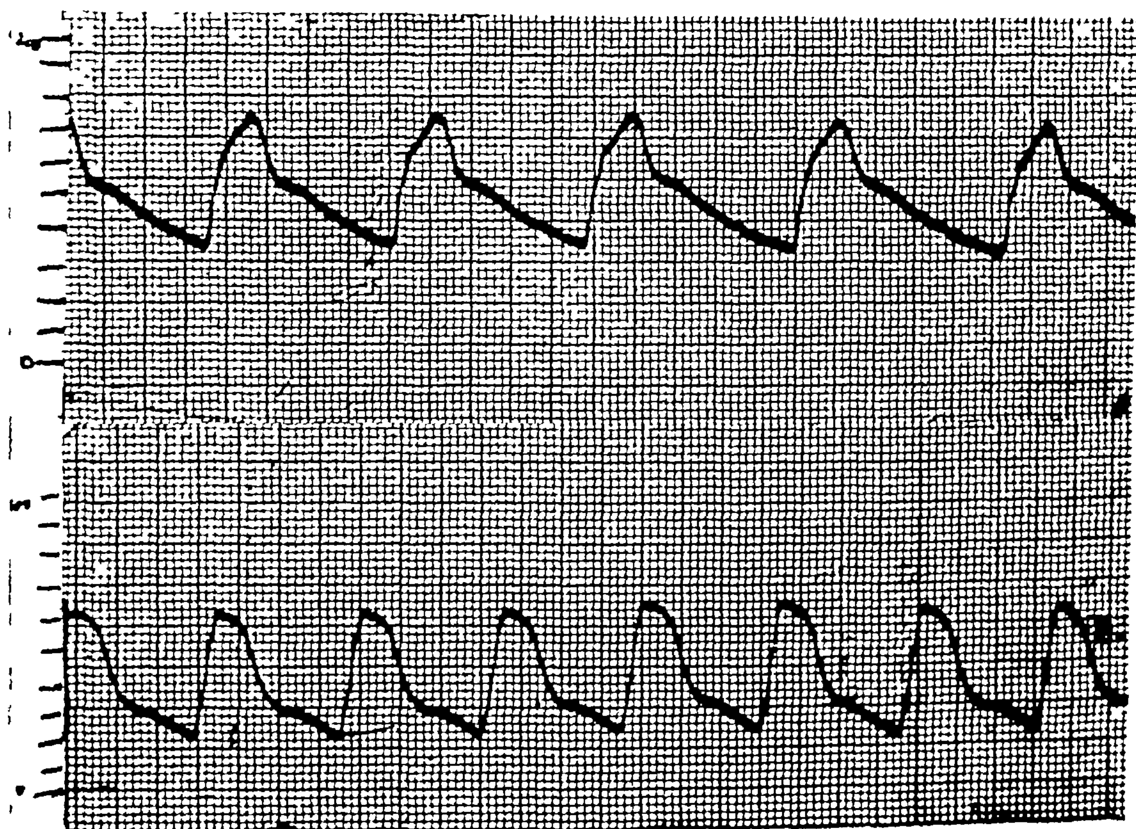


Fig 6B Pre- and postoperative brachial arterial tracings obtained from a patient whose stenosis was relieved by surgery Note the abolishment of the slow rise to a peak

and other evidences of functional coronary insufficiency commonly are reported at this stage Later on, right heart failure may develop with peripheral edema and other symptoms of increased pressure behind the right ventricle.

the aortic valve may be taken either by left heart catheterization, by the posterior paravertebral route (Fig 5A,B,C,-D), or by the use of the bronchoscopic method in which the end of a long needle is introduced through the left

main bronchus into the left atrium. Direct puncture of the left ventricle either through the apex, or by the para xiphoid route, and simultaneous record ing of the brachial arterial pressure will enable one to estimate the gradient The two atrial technics require that a plastic catheter be passed through the needle and then through the mitral valve into the ventricle (The para xiphoid route requires the needle to pass through the right ventricle, through the interventricular septum, and then into the left ventricle.) In a patient with aortic stenosis each of these tech nics is capable of showing a systolic gradient across the aortic valve which, if the obstruction is "significant, will be greater than 30 mm. of mercury by planimetric measurement (an average difference between the aortic and the left ventricular pressures during the different phases of systole) The symp-

toms of angina pectoris in aortic stenosis presumably are caused by a relative disproportion between the amount of work the heart has to do and the amount of oxygenated blood which comes to the left ventricular myocar dium through the coronary arteries which are being perfused at a relatively low pressure head.

A rather simple measurement which may serve as some indication of the presence of severe aortic stenosis is a direct brachial artery pressure tracing which shows a slow increase in pressure after the aortic valve opens, an anacrotic notch on the ascending limb of the pressure curve, or even a double peak (Fig 6A,B C)

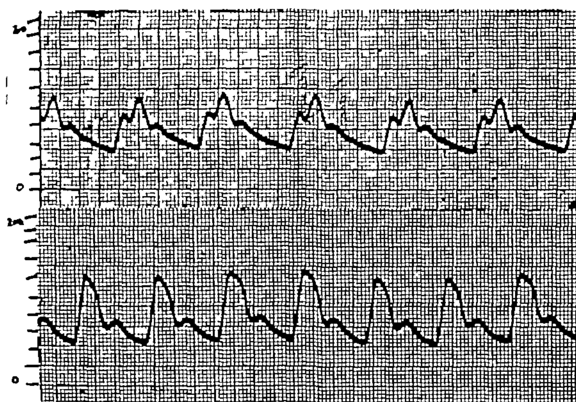


Fig 6C Pre- and postoperative brachial arterial tracing obtained from a patient whose stenosis was relieved by surgery Note the disappearance of the anacrotic notch postoperatively

Symptomatology and Clinical Signs

Symptoms in congenital cases of aortic stenosis may be absent. However

there has been a fairly high incidence of sudden death in these children with no premonitory symptoms In the rheumatic type of aortic stenosis symptoms apparently do not develop until long after the initiation of the described pathologic changes It has commonly been noted that a systolic murmur and thrill may be present for many years in aortic stenosis before the patients develop symptoms Once, however, symptoms do begin, the downhill course is fairly rapid There is a life expectancy of only about two, three, or four years, from the onset of heart failure, syncope, or angina pectoris respectively The most common symptom of aortic stenosis is angina pectoris and the second most frequent is *syncope.* Such fainting attacks may herald sudden death from a bout of ventricular fibrillation The *angina* of aortic stenosis is impossible to differentiate by its character alone from the angina of arteriosclerotic coronary artery disease It may occur on the slightest effort or even at rest as in advanced coronary disease Early, episodes of pulmonary edema and later, chronic ankle edema may characterize the heart failure of aortic stenosis

Physical signs are a late, small pulse (tardus, parvus) which is a function of the small pulse pressure and the low systolic pressure, and the fact that the heart compensates for the obstruction by lengthening the phase of systole as long as possible In severe cases the peripheral arterial blood pressure is low although some patients are seen with moderately hypertensive levels On examination of the heart the apex is found displaced downwards and the cardiac impulse is heaving There is a basal systolic thrill and a murmur is present which often is best heard over the "aortic" area (the first and second interspace) just to the right of the sternum. The thrill and murmur characteristically are transmitted into the carotid arteries The murmur is heard best when the patient leans forward and stops breathing in full expiration Characteristically the murmur is rough, low pitched, and on phonocardiography has a characteristic diamond shape being loudest in midsystole This is in contrast to the murmur of mitral insufficiency which usually is a pansystolic murmur of more even intensity. There may be a systolic aortic ejection "click" which immediately precedes the murmur, and the aortic second sound may be delayed and diminished, or absent in some cases

Chronic atrial fibrillation is seen in less than 10 per cent of the patients with pure aortic stenosis When it does occur, concomitant mitral stenosis and/or insufficiency must be ruled out

On fluoroscopy the left ventricle shows a rounded concentric type of enlargement Calcification of the aortic valve usually will be seen particularly if the patient is over fifty years of age The electrocardiogram consistently shows left ventricular predominance Exceptionally high voltage waves always are seen over the left ventricle and the T-waves may be inverted in these leads In 10 per cent of the cases atrial fibrillation, or varying degrees of atrioventricular block may be found

Diagnosis

The diagnosis of aortic stenosis is made finally only by measuring a significant pressure gradient across the aortic valve In the age group over fifty it is not uncommon to have both a fairly harsh systolic murmur over the aortic valve, apparently due to some thickening, and irregularity in the leaflets, and, at the same time, to have an enlarged heart by physical examination and x-ray study, perhaps because of asso-

ciated coronary artery disease. The absence of a significant gradient across the aortic valve in the presence of a near normal cardiac output distinguishes such patients. It must be stressed that all patients referred for surgery for aortic stenosis should have a direct physiologic determination of the pressure gradient across the aortic valve. Concomitant coronary artery disease may be diagnosed by coronary arteriography injection of the dye being made through a catheter tip placed at the root of the aorta (Fig 7)



Fig 7 Retrograde catheter aortography with injection of 45 cc. of 90 per cent Hypaque now makes possible the visualization of the coronary arteries. In the case illustrated i which temporary cardiac arrest was accomplished with acetylcholine at least two plaques are visible in the left coronary system, as well as almost complete obstruction to the right coronary artery Concomitant successful removal of such plaques by endarterectomy and relief of aortic stenosis by "cuspicure" has been accomplished.

Treatment

The only effective treatment for aortic stenosis is surgical. Some effort at decalcification of the arteriosclerotic valve is being made but has not reached clinical fruition.

Although operations on the aortic valve have been accomplished successfully by closed technics since 1950, it is only since the first open operations utilizing extracorporeal bypass were performed by Lillehei,[1] Bailey [2] and Morrow [3] in 1956 that any really definitive relief of the obstruction could be offered to the majority of the patients. The ineffectiveness of closed technics in the relief of aortic stenosis is perhaps best shown by the lack of consistent relief of the gradient. In addition there has been an operative mortality of 20 per cent for the transventricular dilator method,[4, 5] and an operative mortality of approximately 15 per cent in cases operated on by closed technic utilizing a pouch sutured to the ascending aorta [6, 7]

An entirely open technic through an aortotomy using artificial circulation and antegrade cannulation and perfusion of the coronary arteries to support the myocardium while there is no pressure within the aortic root permits maximal correction of the pathology The choice of procedure then depends upon the pathology Opening the fused commissures is often the least part of the operation

Technic of Relief of Acquired Aortic Stenosis

Anesthesia: The skin is prepared over a forty eight hour period using a *hexachlorophine* preparation (*phisohex*) and covered with towels. The patient is premedicated lightly and is brought to the operating room with at least two large gauge polyethylene catheters inserted into peripheral veins. While breathing pure oxygen *pentothal* is administered rapidly to bring about unconsciousness. *Succinyl choline* (*anectine*) sufficient to produce paralysis is given as an endotracheal tube is introduced. Electrocardiographic monitoring with a cathode ray tube is essential since ventricular fibrillation is not uncommon at this point. The patient is kept "light" throughout the operation, his breathing preferably being controlled by a mechanical ventilator

Thoracic Entrance: A midline skin incision is made from a point one finger above the suprasternal notch to a point approximately 6 cm. beyond the tip of the xiphoid cartilage. In order to obtain an absolutely straight line the skin is previously marked by a ruler or a stretched wire suture

At the same time the chest incision is being made at least one inguinal incision is made to prepare for insertion of monitoring canullae (polyethylene) into the femoral artery and vein If it is intended that a femoral artery is to be used for perfusion return, bilateral inguinal incisions will be made Today we prefer to use the ascending aorta for this purpose

Electrocautery is employed to sear a long line on the anterior surface of the sternal periostium The xiphoid and the suprasternal area are dissected bluntly until the posterior aspect of the sternum is so freed from the mediastinal tissues that a finger or an instrument can be placed underneath the sternum at either end for elevation A special vibrating electric saw is used to divide the sternum in a straight line usually commencing at the lower end The saw produces a smooth even incision without fracture of the tables of the sternum Bone wax is employed to control bleeding from the bone marrow Again the electrocautery is employed to sear the cut edges of the periostium over both the upper and lower surfaces of the sternum A double-bladed sternal spreader is inserted which applies force over separate areas thus tending to obviate fracture of the narrow sternal halves The anterior surface of the pericardium is exposed by carefully dissecting the mediastinal tissue with gauze taking care to avoid rupture into either pleural space The pericardium is opened longitudinally The electrocautery tip is used upon the cut edges of the pericardium to minimize late bleeding after heparinization Extreme care is taken with hemostasis at all times in view of the long period of heparinization which is necessary After the pericardium has been fully opened from its attachment to the aorta above, to the diaphragm below, 3 0 mg of *heparin* per kg body weight is administered The pericardial margins are suspended from the arms of the retractor with heavy sutures placed at one inch intervals along the cut edges

The main pulmonary artery is dissected free of the aorta and encircled with an umbilical tape for later occlusion during total bypass The right atrial appendage is grasped in a noncrushing (Satinsky) clamp and a 3-0 nylon atraumatic purse string suture is applied about its base After eight minutes have elapsed from the time of administration of the heparin a large (#30-40 Fr) catheter is introduced into the right atrium and its multifenestrated tip is brought to rest near the mouth of the inferior vena cava Its blunt malleable obturator-introducer is withdrawn and a pre-bypass baseline blood sample is taken from this catheter before connecting it to the "venous" bypass line The blood is withdrawn from the heart largely by means of gravity (siphon effect) into a plastic reservoir which is situated near the floor (eighteen to thirty-six inches below the right atrium) This reservoir also receives the blood aspirated from the aorta during bypass and is filled with a mass of plastic mesh which is covered with antifoam. In bypass for aortic valve surgery no occlusive tapes are placed around the venae cavae, instead the pulmonary artery is occluded during the period of total bypass by tightening an umbilical tape passed around it

After the catheter has been placed in the right atrium for the venous return and a smaller one has been inserted into a femoral artery or the aorta for the arterial end of the pump, they are attached to the tubing which is led over to the patient preferably from the foot of the operative table Bringing the tubing to the patient in this manner avoids interference with the surgical team and prevents inadvertent obstruction of the flow by one of the members of the team A line for low pressure (nontraumatic) aspiration of blood, and a line with two fine (2 and 3 mm) catheters for coronary perfusion are brought to the field

A heat exchanger forms part of the circuit and the patient's temperature is routinely lowered 10° F. This protects the heart from the temporary anoxia after opening the aorta and before coronary cannulation Also it insures complete adequacy of the bypass in supplying the (thus lowered) needs of the body

The bypass is commenced slowly If there is any evidence of aortic insufficiency the flow rate is kept at a low level (500 cc /min) until the surgical team is ready to cross-clamp the aorta above the valve This prevents "overcoming" the left ventricle and dilating it by the high "diastolic" pressure generated by the pump The intra-arterial pressure monitoring may have been established by means of a small plastic catheter introduced directly into the aortic arch over a needle The arterial pressure wave is visualized on a cathode ray screen and is used as an index of the adequacy of the extracorporeal circulation The mean pressure is maintained at a predetermined level

(usually 70 to 90 mm. Hg) by increasing either the rate of perfusion or the amount of blood in the circuit according to the level of the venous pressure, which is also recorded by similar cannulation of a large vein (perhaps the superior vena cava). The aorta is cross-clamped as high above the valve as is feasible and a special sharp-pointed ("low pressure") suction tube is introduced into the aortic root by stabbing through the intended area of arterial incision. This serves to drain the blood from the heart and lungs which will continue to issue from the valve. The pulmonary artery will have been occluded before cross-clamping the aorta to interrupt the major portion of flow through the lesser circulation. All of

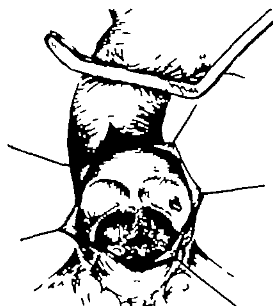


Fig. 8: After cross-clamping the aorta, a vertical incision exposes the valve. (The inferior pole of the incision extends more toward the commissure between the non-coronary-bearing and the right coronary cusp than is shown in the diagram.) (Bailey et al.: The Surgical Treatment of Aortic Stenosis. Encyclopedia of Thoracic Surgery. Courtesy of Springer-Verlag.)

the systemic venous blood returning to the heart thereafter passes extracorporeally by way of the right atrial catheter.

After the heart has become emptied, the incision in the ascending aorta is extended longitudinally taking care to bring the lower end in line with the commissure between the right and the noncoronary aortic valve cusps. The lips of the arterial incision are retracted and the valve is visualized (Fig. 8).

Coronary Perfusion: As quickly as possible each of the two small sponge-rubber cuffed catheters of polyethylene tubing is inserted respectively first into the left coronary ostium, and then into the right. The left is fixed in place by tying down a tiny purse string suture of 3-0 nylon which is placed within the aorta about the ostium. It is tightened, tied, and then wound around the tube and retied. The right coronary catheter after insertion is secured by a simple suture placed from outside the heart around the first portion of the artery. Careful placement of these catheters so the tips do not become blocked, and fixation so that there is no back leakage into the operative field is basic to the success of the entire procedure. As the catheters are inserted coronary perfusion is commenced and is established at a preselected rate of flow by a separate pump which draws from the oxygenator reservoir. The flow rate should be commensurate with the extent of myocardial hypertrophy and may even be as high as 1000 cc. per minute. Usually a flow of 200 to 600 cc. will suffice. During the time it takes to place the coronary catheters and initiate the perfusion the cardiac contraction will become weak and slow and may cease. Some protection of the myocardium during this period is obtained by initially instituting a degree of hypothermia with the heat exchanger. As soon as the coronary perfusion starts, the heart commences to beat again unless ventricular fibrillation has supervened. A special narrow "low pressure" suction tube is introduced through the stenotic valve into the left ventricle to aspirate the bronchial venous return continuously throughout the procedure. This may be withdrawn from time to time if its presence hampers the operator's instrumentation of the valve.

The valve itself is inspected minutely and a feasible plan of attack is formulated. Usually it will become evident that at least one of the cusps can be rendered flexible and mobile by a "sculpturing" technic in which part or all of its calcific encrustation will be removed. A set of special aortic valve rongeurs with biting jaws set at various angles may be employed as well as small aortic valve curettes. Sometimes sharp dissection with pointed scissors will develop a natural cleavage plane (Fig. 9A,B). Great care is taken not to fenestrate one of the valve leaflets by an effort to remove too much tissue at one time. Also great care is taken not to open a commissure which will result in one of the adherent leaflets becoming incompetent. The noncoronary bearing cusp is usually the leaflet which can be best worked upon, and the commissure

between the right and left coronary cusps is usually the most difficult and dangerous commissure to open from the point of view of possibly creating regurgitation The major portion of the operation consists in a careful "manicuring" or sculpturing of the calcium away from the aortic leaflets with the preservation of one of the two endothelialized surfaces of each valve leaflet In most cases the calcific encrustation is most marked upon the concave surfaces of the cusps (Fig 10A,B) One should not attempt to remove encrustations or to "thin" the cusp on both surfaces in the same area lest fenestration result Unfortunately, in some cases it may not be possible to render all the leaflets flexible and the best possible compromise then should be accepted Sometimes one or more of the cusps will be seen to have become "flattened" having lost the normal deep cup-like shape Such cusps cannot support themselves except by virtue of their adherence to a supporting fellow or by virtue of the stiffening which has resulted from scarring and/or calcification Needless to say, such cusps must not be detached from the supporting fellow nor sculptured into flexibility Approximately one hour of meticulous work is necessary on the average to restore a severely stenotic aortic valve to a reasonably satisfactory functional condition This can be obtained by the use of conventional heart-lung bypass plus coronary perfusion as described

Even if the noncoronary cusp is the only one which can be made to become truly flexible, a very satisfactory clinical result may be expected Great care is taken to prevent any small pieces of calcium from falling down into the ventricle and as a preliminary step a strip of packing gauze may be inserted through the opened valve into the ventricular chamber in order to catch any such particles After the valve itself has been as fully decalcified as possible, any subvalvular calcification which may present as masses in the outflow tract should be removed Such subvalvular extension may be particularly prominent immediately below the actual commissures

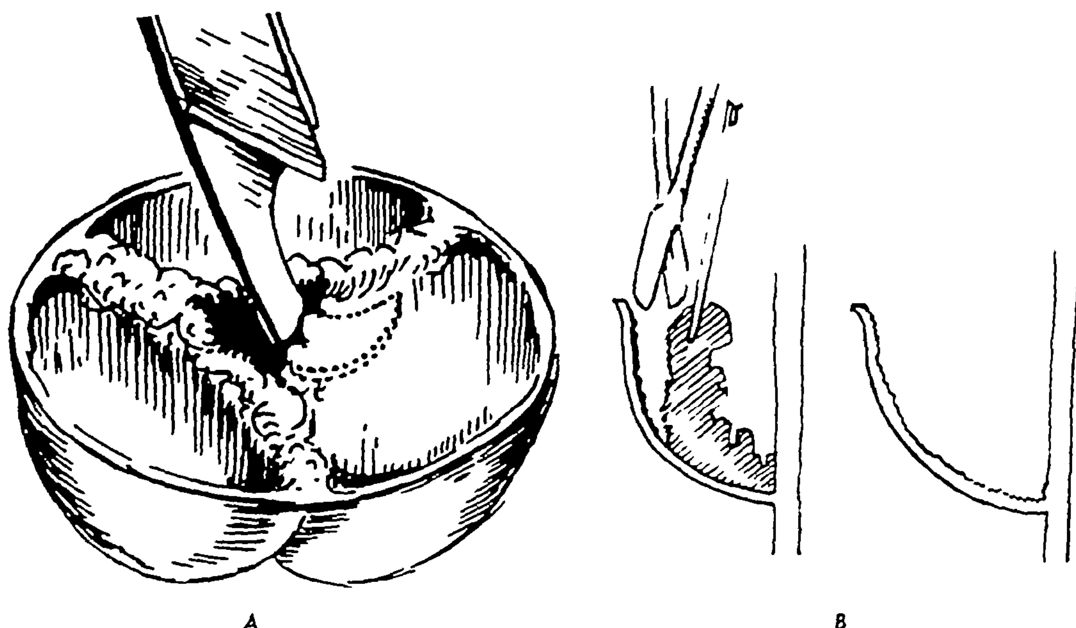


Fig 9 A, The fused calcific commissures may require a special guillotine knife for incision B, The major portion of the operation consists of the gradual removal by rongeurs, scissors (a fine Metzenbaum), and delicate Potts forceps of the calcific material which thickens and stiffens the leaflets

If a fenestration is inadvertently produced by the process of "cuspicure" a small patch of strong plastic material preferably highly compressed *ivalon* reinforced with *teflon* mesh may be applied into the concavity of the cusp to patch-over the perforation

After the valve reconstruction has been completed, the lower half of the aortic incision is repaired very carefully using a continuous everting mattress suture of 4-0 arterial silk, and then an over-and-over suture also of 4-0 silk Particular care must be taken in the closure of the lower end of the aortic incision

since after the heart has assumed the burden of maintaining the circulation it becomes extremely difficult to work in this area.

A critical moment occurs at the time the heart is taken off the coronary perfusion and placed on its own (aortic) perfusion which should be accomplished quickly to avoid the development of myocardial anoxia If the ventricles are fibrillating, coordinated contractions should be restored before interrupting the coronary perfusion. In rapid succession the sutures holding the coronary perfusion less than the total estimated flow for the patient in order to avoid "over-coming" the heart by the high maintained "diastolic" or mean aortic pressure. The venous pressure and arterial pressure levels must be watched very carefully at this point to make sure that the proper amount of blood is circulating both through the body and the pump The arterial wound is sutured and the clamp is removed. As the patient's heart gives evidence of its ability to take over more and more of the circulatory burden, the pump output is



Fig 10: A, Typical aortic valve with severe calcification distorting all three leaflets, and completely obliterating the commissure to the right (The ostia of the left coronary artery is visible in A and B to the lower right side of the valve in the everted aortic wall.) B The same valve after meticulous "sculpturing" or cuspicore of the leaflets. The left coronary cusp is now restored to a near-normal thickness with resultant flexibility and mobility There is no loss of competence inasmuch as the deep pocket-like structure of the cusp has been regained without perforation. (Bailey C. P., Musser B. G and Morse, D P.: Cardiology 4 147 1959)

catheters are cut loose and these catheters and sutures are removed. The heart is allowed to fill with blood by releasing the tourniquet about the pulmonary artery and by partially blocking the venous drainage from the right atrial catheter As soon as blood flows freely from the aortic incision, having displaced all entrapped air from the ventricle and aorta, the remaining aortic incision is clamped shut using a special dentate (Beck) clamp At the same moment the aortic cross-clamp is removed. Immediately thereafter the output of the pump oxygenator is reduced to somewhat reduced correspondingly Finally the perfusion is terminated, and if the circulatory status remains satisfactory decannulation is begun. The catheters are removed first from the atrium, and then from the artery and the heparin is neutralized with polybrene in a ratio of 1 1 mg The margins of the thoracic wound are examined for bleeding points and additional sutures are placed if required The arterial wounds are repaired with fine arterial silk. The atrial appendage is tied, and the cut end is oversewn with atraumatic silk sutures. Finally the pericardium is closed with inter

Bailey C. P., Musser B G., and Morse, D P Am. J Cardiol., 4 147 1959
3 Morrow A. G.: Quoted in personal communication from W J Zukel (Public Health Service) to C. P Bailey June 1956.
4 Bailey C. P., Ramirez, H P., and Larzelere, H B., J.A.M.A., 150·1647 1952.
5 Bailey C. P., Bolton, H E., Jamison, W L., and Nichols, H T Circulation, 9·22, 1954
6. Bailey C. P., Surgery of the Heart. Lea & Febiger Philadelphia, 1955
7 Bailey C. P., Bolton, H E., Nichols, H T., and Likoff W Encyclopedia of Thoracic Surgery E. Derra, Editor Springer Verlag Berlin, 1959 Vol II, p 866.

SURGERY FOR AORTIC INSUFFICIENCY

Jacob Zimmerman, F.R.C.S., and Charles P. Bailey, M.D.

The clinical and physiologic entity known as aortic regurgitation is common to several varieties of anatomic deformity of the aortic valve. True corrective surgical therapy for incompetence of this valve, therefore, must be adaptable to each of these different pathologic pictures. Certain recent advances in surgery, particularly total bodily perfusion, have opened the way to direct corrective procedures for most types of aortic valvular incompetence. This condition, for which only a decade ago surgical treatment was practically inconceivable and which five years ago was occasionally given to indirect surgical alleviation,[1-4] has become amenable to operative correction to the extent that surgical cure now may be offered to many patients and considerable prolongation of life and improvement in health can be provided to the great majority of sufferers.[5, 6]

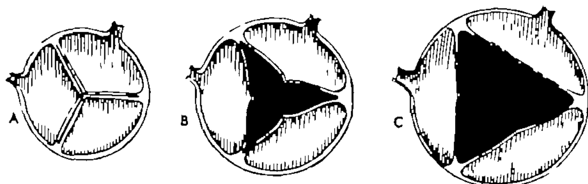


Fig. 1. Development of aortic incompetence due to enlargement or overstretch of the aortic root. A. Relationship of a normal valve within a normal sized aorta. B. Although the valve cusps may remain individually normal, sufficiently marked enlargement of the aortic root will prevent diastolic coaptation of their margins. C. When the aorta becomes extremely dilated the free margins of the three cusps will be stretched like bowstrings across 120° arcs of the aortic circumference.

Pathology

Common to all types of lesions resulting in aortic regurgitation is the fact that the available valve tissue is incapable of guarding the entire valve area during cardiac diastole. Among these lesions are perforations of a cusp, an actual dearth of valve tissue perhaps due to shrinkage and retraction, prolapse of a cusp, laceration of one of the cusps, or circumferential widening of the aortic root which will not permit the otherwise structurally normal cusps to coapt effectively. Although certain pathologic pictures tend to be associated with certain etiologic factors, this relationship is not always consistent.

The pathologic types most commonly met with are four:

I. Circumferential Widening of Aortic Root: The classical etiologic factor for this type of deformity is luetic aortitis. However, similar incompetence may result from rheumatic weakening of the aortic root, Marfan's disease, and perhaps congenital weakness of the aortic wall (Fig. 1A,B,C). In the typical case, the valve cusps are relatively normal in texture, size, and contour although their free margins may come to be stretched like bow strings each across one third of the circumference of the

dilated aorta Therefore, they will have been rendered completely incapable of fulfilling their coaptive function Not infrequently, however, the cusps may become involved secondarily either through the traumatic effect of the regurgitating blood stream or in response to additional pathologic factors In most cases the circumferential widening is due to direct involvement of the aortic wall itself without any other specific evidence either of luetic involvement of the root of the aorta or of

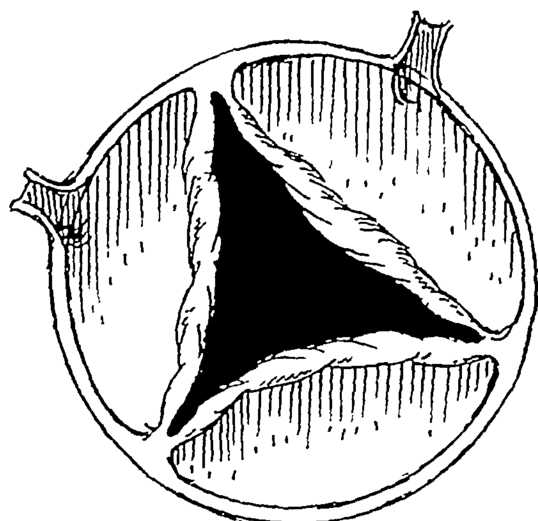


Fig 2 Rheumatic aortic valvulitis frequently results in eversion or retraction of the margins of the cusps so that coaptation cannot take place

rheumatic involvement of the valve cusps

II. Cusp Deformation: By far the commonest lesions (at least in our experience) involve the leaflets themselves In the majority of cases the underlying etiologic factor is the rheumatic process which produces thickening and shortening of the cusps and retraction of their edges which frequently may be seen rolled towards the aortic circumference (Fig 2). Usually not all of the cusps are affected to the same degree. However, a severe involvement of merely one of them may result in a very marked degree of valvular insufficiency

Occasionally the valvular incompetence is the result not of retraction and shortening of the cusp edge but rather of its pathologic elongation Such elongation of the valve edge (probably the result of the repetitive traumatic effect of the diastolic press or reflux upon a pathologically weakened valve edge) causes the involved cusp to prolapse in the

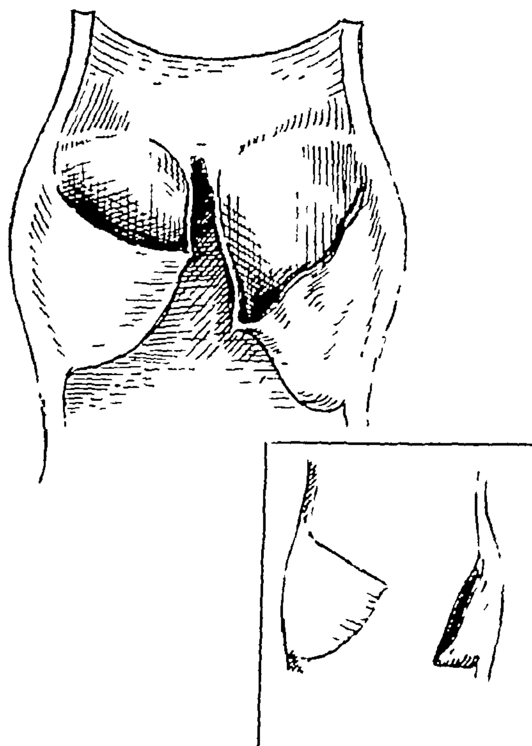
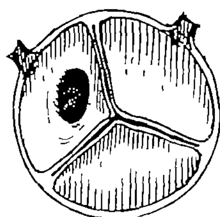


Fig 3 The normal aortic valve cusp when distended with blood assumes a contour not unlike that of the female breast Extreme elongation of the free margin of a cusp, retraction of the central portion of a cusp, or attachment (congenital) to less than one third of the aortic circumference may lead to prolapse and incompetence (see inset)

direction of the left ventricular outflow tract during diastole. Because of this prolapse, the leaflet edge falls below those of its fellows and no effective coaptation of the cusps at the natural valve level is possible (Fig 3). Occasionally such a prolapsing cusp is part of a congenital deformity of the valve, not uncommonly seen in conjunction with a "high" ventricular septal defect or a congenitally bicuspid aortic valve Thickening and hardening of the valve



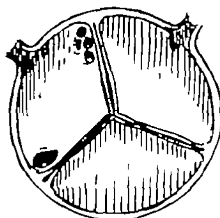


Fig 4 Perforation (fenestratio) of the aortic valve cusps. A. Subacute or acute bacterial infection may produce a large perforation of the center of a cusp. B In congenital fenestration one or more small points of interrupted continuity of a cusp may be see usually in close relationship to one of the commissures. Because they tend to be buttressed by the adjacent leaflet during diastole they do not necessarily cause incompetence. Sometimes several small defects will become coalesced to form a single larger one which then may become hydrodynamically significant

tissue in itself which is seen characteristically in rheumatic lesions, may have a deleterious effect upon valvular movement and, in this way impair valvular competence.

Fenestration of one or more cusps may be the result of congenital malformation or may be caused by bacterial endocarditis or trauma, usually surgical (Fig 4A B)

III. Incompetent Valve with Stenosis: The pathologic aortic valve has a marked predilection for calcification and a propensity toward the development of superimposed bacterial infection The reasons for this are obscure but probably are related to the severe and incessant trauma to which this valve is subjected in its natural functioning As a result of one of several different etiologic factors, most commonly the rheumatic process, the aortic valve cusps progressively may become fused together in a centripetal fashion and the whole structure may become converted into a calcified shelf This is the usual pathologic picture in severe aortic stenosis. Because of the fixation of the valve cusps, if the commissural obliteration is symmetrical, a certain degree of valvular incompetence necessarily must be present (Fig 5) When the cross fusion of the leaflets is assymmetric one



Fig. 5 Classical deformity in symmetric aortic stenosis. Note the partially obliterated commissures, the eversion of the margins of the cusps, and the calcific incrustations upon the concave aspects of the cusps. Since the hardened leaflets co not coapt f lly during diastole some degree of incompetence necessarily coexists with symmetric stenosis. (Larzelere H B., and Bailey C. P., J Thoracic Surg., 26:31 (July) 1953.)

Signs and Symptoms

The clinical picture of aortic insufficiency is most profoundly influenced by the condition of the left ventricular myocardium As a result of its remarkable compensatory powers, unless the regurgitation is excessive from the very beginning, the patient usually is asymptomatic during the earliest stage of the disease However, the typical early blowing diastolic murmur will be heard, usually best over the aortic area and transmitted downward along the left sternal border Left ventricular strain may be revealed by the electrocardiogram Frequently the diastolic blood pressure is lower than normal, resulting in a comparatively wide pulse pressure.

This asymptomatic state sometimes may last for 10 to 20 years or more and may throw a cloak of benignity over a basically malignant type of cardiac dysfunction However, there are other cases in which the entire course is measured in weeks or months

At a more intermediate stage, the clinical picture is governed by the circulatory capacity of the considerably dilated and hypertrophied left ventricle Apart from certain limitations at times of excessive exertion, the patient may not be obviously symptomatic A common distressing symptom is the sensation of the powerful heart beat which may disturb the patient's rest and sleep. The pulse pressure at this stage may be very wide, frequently with a high systolic as well as a low diastolic level The high systolic pressure probably is caused chiefly by an increase in the amount of blood ejected with each contraction of the ventricle The low diastolic pressure is related, in part, to the lack of diastolic restraint at the valve level and also, in part, to a "compensatory" peripheral vasodilatation Teleologically this latter feature might be looked upon as helpful to the forward flow of the blood to the tissues Combinations of these two factors produce the classical "water hammer" pulse as well as Corrigan's sign The wide pulse pressure frequently is especially marked in the lower extremities, the systolic pressure in the legs in these cases being elevated out of proportion to that in the arms

The terminal stage of the disease starts when the left ventricle has reached the limits of its compensatory powers Clinically the failure of the heart when limited to the left ventricle is expressed in an anginal syndrome upon exertion, in bouts of syncope, and in episodes of acute pulmonary edema When not so limited signs and symptoms of complete congestive heart failure are not uncommon and are of the most serious prognostic significance. At this stage the life of the patient is in grave immediate danger since the heart (which frequently is of considerable size) is in an extremely precarious condition often evidenced by electrocardiographic signs of left ventricular irritability Episodes of precordial pain at rest, which are indicative of a state of severe coronary insufficiency, are not uncommon. Death may come suddenly with or without severe exertion The patient, although he may not appear seriously ill on the surface, should be considered but a mere human shell with core so deteriorated that it may crumble at any time

Diagnosis

The diagnosis of aortic regurgitation is comparatively easily made. It depends primarily upon the existence of the typical diastolic murmur best heard over the aortic base and transmitted down the left border of the sternum.

The murmur is heard early in diastole, is decrescendo and blowing in character. Frequently a systolic component is present. This does not necessarily indicate the existence of an element of stenosis.

However aortic stenosis may coexist with insufficiency and then may even be the predominant lesion. A certain amount of regurgitation, probably functionally insignificant, is a very common concomitant of aortic stenosis. Associated mitral stenosis, too is not infrequent although the mere fact that a diastolic murmur is audible at the apex does not necessarily indicate the existence of mitral obstruction. Such a finding when present as an isolated one without other evidence of mitral stenosis (such as an enlarged left atrium) may represent the so-called Austin Flint murmur.

Because of the possible presence of other valvular lesions, the importance of which may be difficult to evaluate, it often will be necessary to perform a number of special diagnostic studies.

Left heart catheterization through a left atrial puncture generally is indicated in the presence of left atrial enlargement. It will help to clarify whether or not mitral stenosis is present and to indicate its severity. The same study by advancement of the catheter often also will detect the presence of a significant component of aortic stenosis by measurement of a transaortic gradient. Or a gradient between the left ventricular and the brachial arterial systolic pressures may be established by simultaneous direct needle puncture of the artery after advancement of the catheter tip into the ventricle.

In patients with a normal sized left atrium and in whom no suspicion of mitral stenosis exists the gradient across the aortic valve can be measured roughly by means of simultaneous left ventricular and brachial arterial puncture. The left ventricular puncture also will permit the simultaneous performance of a left ventriculogram which will determine the presence or absence of mitral insufficiency. It is, however clinically rare to find a severe degree of mitral regurgitation in the presence of marked aortic valvular disease. Evidently the existence of the former lesion tends to preclude the generation of sufficient ejection power within the left ventricle for such a patient to survive very long.

In addition to the qualitative diagnosis of aortic regurgitation and the evaluation of any concomitant valvular lesion two other aspects of the diagnosis are important. The quantitation of the magnitude of the aortic regurgitation is particularly important during the first and second stages of the disease while the clinical condition still is good. A good quantitative study might help to bring the prognosis of the individual patient into better perspective. The size of the pulse pressure and the decreased level of the diastolic peripheral blood pressure offer little guidance in quantitation of the leak especially in the presence of acute or chronic left ventricular failure. The most reliable although admittedly still rough method of quantitation consists of serial radiographic study following injection of a radiopaque substance into the ascending aorta (aortography). This may be carried out either by percutaneous (suprasternal) needle puncture or by catheterization of a peripheral artery (preferably the femoral). The aortic regurgitation then can be judged radiographically in grades of one to four plus in accordance with the observed degree of opacification of the left ventricle.

Of special interest to the surgeon is the anatomic diagnosis of the type of lesion A careful evaluation of the clinical picture can be helpful in this direction The history of rheumatic fever or the coexistence of mitral or aortic stenosis suggests the pathologic lesions described as Types II or III The absence of a prominent harsh systolic murmur and violent expansile pulsation of the ascending aorta on fluoroscopy suggest a Type I or Type II deformity Any indications of the previous presence of lues are suggestive of a Type I lesion A history of sudden development of typical signs and symptoms of aortic regurgitation following blunt trauma to the thorax, coincident with operative trauma to the aortic valve (surgery for aortic stenosis), or occurring during the course of bacterial endocarditis suggests a Type IV lesion.

Cineradiographic study during opacification of the ascending aorta may be helpful in this respect since it may show an eccentric jet of regurgitation or an abnormally wide ascending aorta suggesting circumferential widening of the aortic root (Fig 21A)

In the great majority of patients, however, the exact anatomic diagnosis can be made only by inspection of the valve at the operating table

Treatment

Until the advent of true intracardiac surgery 12 years ago, no effective definitive treatment for aortic regurgitation was conceivable In view of the practically asymptomatic status of the disease in its earlier stages, there did not appear to exist any need for treatment in this phase. The left ventricle seemed to be capable of taking care of the situation very successfully. Only with the development of failure, left-sided or congestive, did the physician enter the clinical picture therapeutically. Unfortunately, at this stage of the disease, medical measures are of very limited value and heart failure is but temporarily controlled or held off

Due to the very serious associated mortality and the basic ineffectiveness of early operative attempts at the correction of aortic insufficiency[1, 2] the philosophy underlying the application of surgical therapy was somewhat similar to the classical medical attitude just described. Surgical intervention was called for only as a last resort when the patient's imminent demise was well in sight The placement of an artificial valve mechanism into the descending aorta[3, 4] for a time did gain a certain degree of popularity Certain fundamental defects of this method together with the great strides which have been made in the field of "open-heart" surgery during the last few years have combined to relegate the Hufnagel valve technic to medical history.

During the past three years definitive surgical treatment for aortic insufficiency has become a reality For most of these patients surgical treatment during the first and second stages of the disease offers the opportunity for either complete correction or, at least, very marked improvement of the condition Unhappily, however, many physicians still look upon the earlier stages of the disease as basically benign and many patients still reach the surgeon during the very last stages of decompensation It must be obvious that in such patients surgical procedures which involve an "open-heart" technic necessarily carry very serious additional risks due to the extremely poor condition of the left ventricle upon the integrity of which the survival and recovery of the patient must depend

Earlier operation as a result of more

complete understanding of the natural history of aortic regurgitation, early investigation by quantitation, and a more general appreciation of the possibilities of the surgical technics available will *tend, in the future, to reduce the operative mortality considerably* and will offer bright new hope to many sufferers from this disease.

Surgical Considerations and Concepts

The potentialities and limitations of surgical therapy for any lesion are most profoundly influenced by the anatomic, physiologic, and pathologic factors involved. Probably in no other region in the body are these factors basically as unfavorable to the surgeon as in aortic valve disease. The position of the aortic valve within the root of the aorta in the very depth of the heart, its primary functional importance for moment to-moment survival, together with the impossibility of predicting from preoperative studies the exact type of pathology involved have posed until recently practically insurmountable obstacles to effective correction of aortic insufficiency

Only since the advent of "open heart" surgical technics could serious definitive attempts at the correction of aortic incompetence be considered. The apparent need for interruption of the coronary circulation during open aortotomy however has proffered a serious limitation to feasible technical accomplishment because of necessary restriction of the permissible operative time. Elective cardiac arrest for the purpose of significantly reducing cardiac metabolism has proved disappointing The myocardium and especially the intracardiac conduction tissue is very sensitive to oxygen lack, particularly when hypertrophy of the muscle is present. The oxygen requirements are basically the same for the heart while under elective arrest as when beating without resistance.[8] The solution of this problem was provided by the innovation of technics for artificial coronary perfusion with special catheters which can be placed and temporarily fixed within the coronary ostia. Recently it has been shown that rather high coronary perfusion rates (500 to 800 cc. for the average patient) are preferable. Although the conventional pump-oxygenator bypass technics, with the addition of coronary perfusion, provide reasonably satisfactory basic conditions for the performance of definitive corrective procedures upon the aortic valve, certain modified bypass technics permit longer and safer periods for the actual operative work

The autogenous-lung bypass system[9] which provides temporary replacement of cardiac function only (in contradistinction to standard pump-oxygenator technics which replace both heart and lung functions) results in fewer deleterious effects upon the blood and therefore can be used for much longer periods of bypass. More recently it has been found in practice that the combination of the somewhat simpler conventional bypass system with the induction of bodily hypothermia (using heat exchangers interposed into the bypass system) and direct coronary perfusion safely provides a sufficiently prolonged operative time for most surgical procedures upon the aortic valve.

The concepts underlying the surgical correction of aortic valvular incompetence are based both upon certain specific aspects of the normal anatomy of the aortic root and upon the type of pathology found

The root of the aorta which is attached to the anterior aspect of the

combined inflow-outflow aperture of the left ventricle is best visualized as being anchored to the myocardium by the three low points of its cusp attachment[10] (Fig 9) At two of the points the attachment is to strong fibrous bodies (the right and left fibrous trigones) Anteriorly, however, the aortic root is attached directly to the upper (cephalic) edge of the ventricular septum It is continuous with the conus ligament through which the pulmonary valve is attached to the aortic valve Close to the junction of the low point

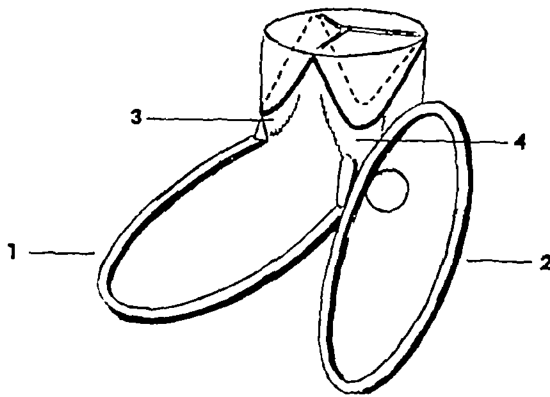


Fig 9 Attachments of aortic valve cusps and their relationships to the fibrous structures which make up the skeleton of the heart (seen from the atrial aspect) (1) Annulus fibrosus of the mitral valve Note the "horseshoe" shape of this structure (2) Annulus fibrosus of the tricuspid valve (3, 4) Attachments of the vortices of the left and the noncoronary aortic valve cusps to the left and right fibrous trigones, respectively

of the noncoronary bearing cusp with the right fibrous trigone the common conduction bundle pierces the fibrous ring of the tricuspid valve to reach the ventricular septum at which point it divides into its right and left branches, the latter piercing the septum to run down along its left ventricular surface as a group of discrete branches

It is important to realize that the standard textbook description of a circular annulus fibrosus of the aortic valve cannot be demonstrated in the fresh heart The only structure at the root of the aorta which can be utilized surgically as an "equivalent" of an annulus fibrosus is the zigzag line of fibrous condensation which marks the attachment of the three cusps to the aortic wall (Fig 10).

The aortic valve mechanism consists of three cup-like cusps arising from the aortic wall which obstruct retrograde blood flow during diastole by moving

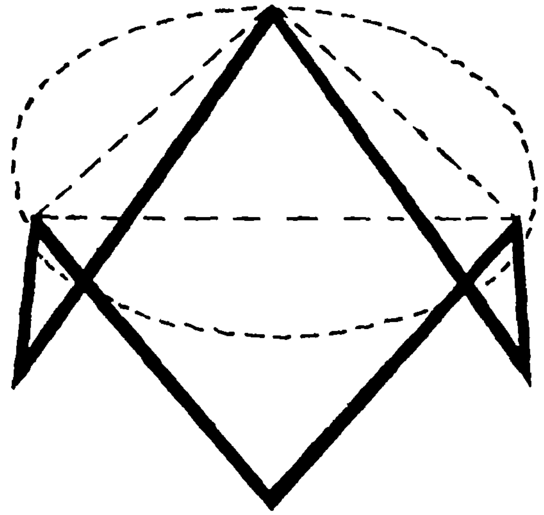


Fig 10 Zig zag line of fibrous condensation which marks the attachment of the individual aortic cusps to the arterial wall Note that there is no other fibrous structure in this region which might be designated an aortic annulus fibrosus

centripetally to mutual apposition as they fill with blood Competence of the valve is dependent upon uninterrupted contact of the three cusps along their free margins and over a narrow zone of medial contact (lunulae and corpora arantii) This mechanism provides a very wide opening during ejection since the three cusps move in three different directions to evade the "bolus" of ejection The competence of such a delicate and precise mechanism, however, is dependent upon the continued anatomical and functional integrity of all three cusps and, therefore, is comparatively easily deranged

Restitution of competence to the valve mechanism in its tricuspid form so

far has proven impractical in most cases except in those in which the regurgitation is due to perforation of a cusp proper the rest of the mechanism remaining unchanged. When the incompetence is caused by a disturbed relationship between the cusps whether due to their intrinsic pathologic changes or to circumferential widening of the aortic root, it usually has been found best to restore valvular competence by converting the tricuspid valve structure into a bicuspid one, or even a unicuspid one.[5 6] This concept aims at the creation of a valve mechanism which is much simpler and far less delicate. Valvular competence thereafter depends upon the continuous contact of two cusps or similar structures along a single line of contact. It cannot be denied that such a mechanism is not as ideal as the tricuspid one, since the effective opening during systole is not as wide as would be present in the latter instance (Fig 11A,B C,D) However the resultant reduction in flow area, in practice, is not too serious and must be considered a price well worth paying for the restitution of valvular competence.

Operative Procedures

Extracorporeal Bypass: The bypass system which we use most commonly for aortic surgery is that shown in Figure 12 A single heat exchanger may be utilized or separate ones may be introduced into both the "venous" and the "arterial" lines of the extracorporeal circuit in order to combine the advantages of readily controllable bodily hypothermia.

Incision and Cannulation A long midline sternum splitting incision and the following cannulation technic is employed The pericardium is incised longitudinally and after general bodily heparinization by intravenous injection (3 0 mg heparin sulfate per kilogram of body weight) the pulmonary artery is dissected free from the aortic root and is encircled with a tape (Fig 13A,B) Should an injury of the pulmonary arterial wall inadvertently be produced during the dissection it may be sutured directly with fine arterial silk. Should this be impracticable, because of inaccessibility of the perforation, the site of dissection may be packed with oxidized cellulose and the ventral margins of the area sutured together Very adequate control of the situation is provided in this way The end of the right atrial appendage is circumscribed with a purse string suture of fine but strong nylon thread (double or triple zero) and is clamped proximally An incision is made within the purse stringed area and the multi ply perforated end of a large (#38-42 Fr) plastic catheter stiffened with a Fitch[11] malleable obturator is inserted as the clamp is released. The end is positioned within the mouth of the inferior vena cava. The long ends of the purse string suture are tied, then are passed twice about the catheter and are retied. The obturator is removed and the catheter is attached securely to the venous tubing of the extracorporeal system (Fig 14) Any gross quantity of contained air should be excluded from the lines while making this connection.

While one or both of the femoral arteries may be utilized for the return of the oxygenated blood to the body we often have preferred to use the distal ascending aorta since this technic permits the use of a much larger catheter This is of especial importance when a pulsatile flow pump is used since less blood can be moved through a tube of given caliber with a pulsatile than with a continuous-flow mechanism

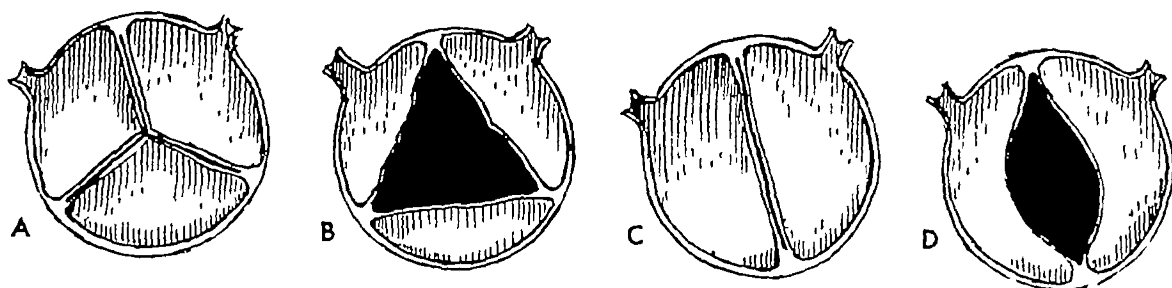


Fig 11 Diagrams which illustrate the wider functional orifice which is provided by a tricuspid valve mechanism than by a bicuspid one However, the simpler bicuspid valve is more certain to close and structurally provides a stronger defense against regurgitation *A* Normal tricuspid aortic valve structure, closed *B* Full functional opening afforded by a tricuspid type of valve (Da Vinci, MacMillan) C Bicuspid valve, closed *D* Bicuspid valve, open Note how much smaller this functional orifice is than that shown in 11B

Fig 12 Usual extracorporeal circuit employed by the authors for open aortic valve surgery *A* Single atrial catheter, multifenestrated *B* Venous reservoir C Heat exchanger on the venous side *D* Jehle oxygenator *E* Heat exchanger on the arterial side *F* Pulsatile flow pump G Filter bubble-trap *H* Arterial catheter inserted directly into the ascending aorta

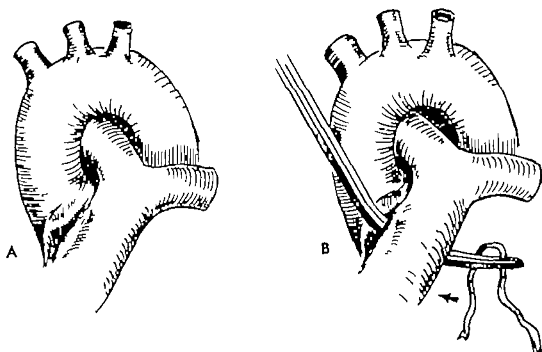


Fig. 13 Tape encirclement of the pulmonary artery A. Limited dissection of the area of adherence between the aorta and the main pulmonary artery B. Once an instrument can be guided between the two arteries an umbilical tape is drawn between them.

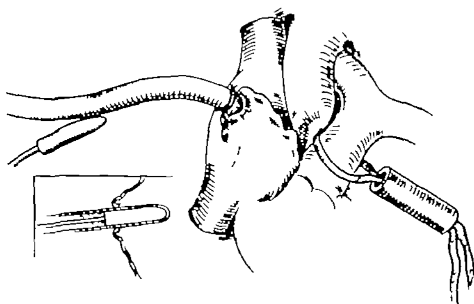


Fig. 14: Method of insertion of the atrial catheter utilizing a malleable obturator to lessen momentary bleeding (inset) Note the method of applying tourniquet constriction to the encircled pulmonary artery

We usually expose and catheterize (with fine polyvinyl tubing) both the right femoral artery and the right saphenous vein for electronic pressure monitoring during the course of the operation. Care should be taken not to restrict the amount of aortic wall available for autogenous grafting

An oval area including the upper two centimeters of the anterior aspect of the ascending aorta is purse-stringed with strong fine nylon suture (two zero) an attempt being made to avoid piercing the lumen by passing the swedged-on needle only within the thickness of the aortic wall (subadventitially). This area is excluded from rest of the aorta by a dentate clamp (Beck aortic) and a longitudinal incision is made in the excluded portion which should be just large enough to admit the preselected aortic catheter.

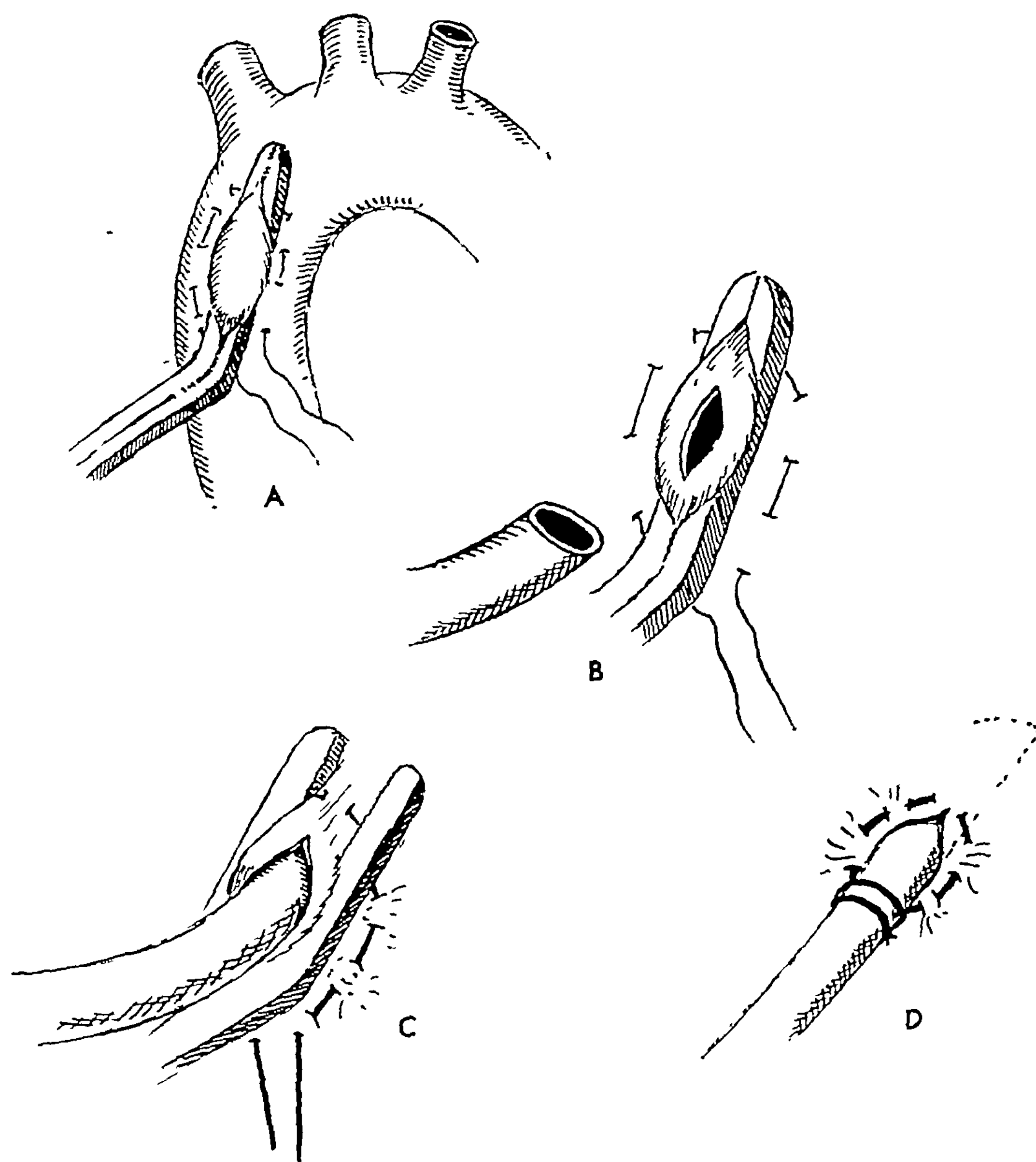


Fig 15 Method of cannulating the cephalic portion of the ascending aorta *A* After encircling the selected site of entrance with a purse string suture of 2 zero nylon thread this area is excluded with a noncrushing clamp *B* The excluded area is incised longitudinally just far enough to admit the preselected catheter *C* As the clamp is released the catheter is advanced and the purse-string is tightened *D* The ends of the tied purse-string suture are passed twice about the catheter, then retied This maneuver should be repeated once

The latter (usually 32-40 Fr) is cut to a length of 20 cm., is beveled and rounded terminally with scissors and is stiffened with the appropriate sized semimalleable (Fitch) obturator As the clamp is released the beveled end of the tube is passed through the aortic incision and advanced to the beginning of the descending aorta The purse string suture is tightened and tied. The long ends are each passed around the tube and tied this maneuver is then repeated. The obturator is removed and the catheter is secured to the "arterial line of the bypass, extreme care being taken to exclude all air from this part of the system (Fig 15A,B C, D)

Partial Bypass: At a given signal the obstructing clamps are removed from the arterial and venous lines and the pump is started at a low rate of flow This is increased gradually to a pre selected but subtotal level. This rate must be so adjusted that the left ven tricle does not become overdistended by diastolic filling through the incompetent aortic valve. With the initiation of the bypass, cooling of the blood may be accomplished by flowing cold water through the heat exchanger The bodily temperature is monitored by thermometers or thermocouples placed within the esophagus, the pericardium, or one of the venae cavae When it reaches the preselected level (usually from 25 to 30 C.) cooling is ter minated and the flow rate of the perfusion is adjusted to the calculated "total" level (usually from 50 to 70 cc. per kilogram of body weight at normothermic temperatures, appropriately less at hypothermic levels) The tape about the pulmonary artery is tight ened establishing total bypass.

Aortotomy At a given signal the ascending aorta is cross-clamped, a longitudinal stab wound is made in the anterior aspect of the aortic root, and a "low pressure" suction device is in serted into the region of the aortic valve. The initial profuse pulsatile flow of blood soon ceases and the aortic stab wound is extended in curvilinear



Fig 16: Exposure of the aortic valve. After the bypass is complete the supravalvular portion of the aorta is incised longitudinally as it is cross-clamped at a higher level. The cardiac extremity of the somewhat curvilinear incision is made to terminate over the commissure between the right coronary and the noncoronary cusps.

fashion upward nearly to the cross-clamp and downward to the aortic valve level. The exact direction of the lower extension is guided by intermit tent visualization of the valve so that the incision terminates exactly over the commissure between the noncoronary and the right coronary valve cusps (Fig 16) The "low pressure" suction tip is passed through the valve into the outflow tract of the left ventricle.

Coronary Cannulation and Perfusion: Both Kay[13] and our group[14] independently and nearly simultaneously recognized the desirability of perfusing the coronary arteries during open aortic surgery in order to maintain the viability of the myocardium The principle was first applied in a human patient on April 18, 1958[14]

The ostium of the left coronary artery is visualized and a fine nylon suture (3-0) swedged upon a small curved needle is placed about it in purse-string fashion taking fairly deep

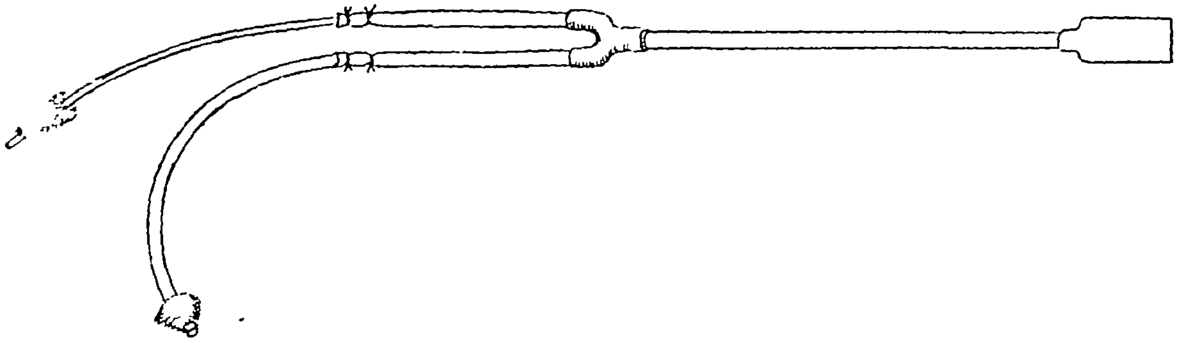


Fig 17A Sponge rubber cuffed (Nunez-Tello) polyethylene catheters for individual cannulation of the coronary arteries (Available from the Abbott Laboratories, Chicago, Ill)

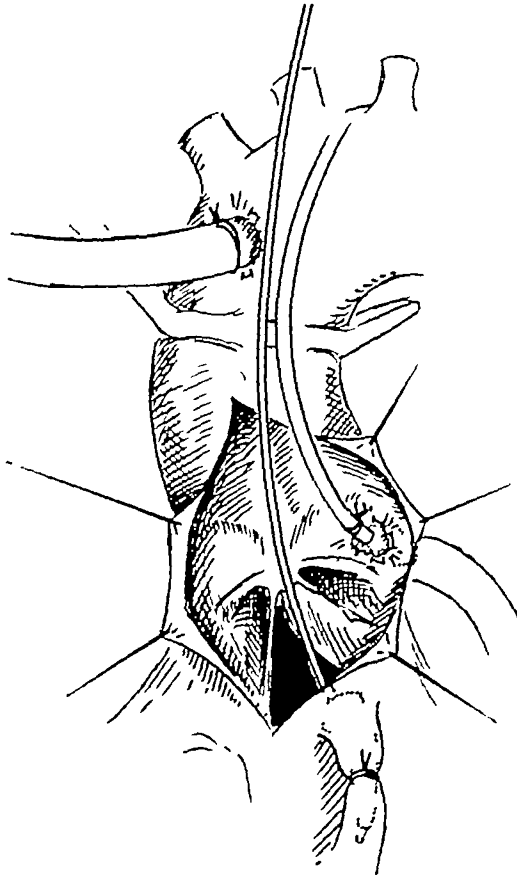


Fig 17B Method of placement and suture fixation of the coronary catheters

bites into the aortic wall. The sponge rubber cuffed end of a Nunez Tello[12] left coronary catheter which already has been connected to the extracorporeal system and is filled with blood is passed well into the lumen of the left coronary artery. The ends of the tiny purse string are tightened and tied, and then are passed about the catheter and re tied. Coronary perfusion is begun, a flow of the order of 500 cc. per minute being used at a body temperature of 37 C, appropriately less is required at lower temperatures.

While a similar technic may be used for cannulation of the right coronary artery, in practice it often has been simpler merely to insert the rounded end of a suitably bent hollow metal tube directly into the ostium. This already has been joined to the other side of a Y-connection in the coronary perfusion line and is then hand held by an assistant, thus serving as a retractor for the aortic incision as well as a conductor of blood (Fig 17A,B). Now the total coronary perfusion flow rate is stepped up by 50 per cent.

Closure of the Aorta: After completion of the definitive surgery the lower and upper thirds of the aortic opening are closed by placement of two rows of fine silk sutures (one mattress and one continuous). Then the left coronary catheter is released by cutting and removing the affixing purse string suture. This line is clamped and the coronary perfusion is terminated. The right coronary (metal) cannula is taken out, and the tape about the pulmonary artery is released permitting restoration of a portion of the natural mechanism of perfusion of the lungs by right ventricular contraction.

A dentate clamp (Beck aortic) is placed in such a way that its closure will approximate the lips of the remaining aortic opening. these lips are elevated by traction upon the long ends of the previously placed reparative sutures. The rate of pumping is slowed by 50 per cent and the operator compresses the left ventricle vigorously several times in order to expel any entrapped air. An assistant closes the dentate clamp at the height of one of these episodes of expulsion of blood from the aorta. The aortic cross-clamp is released immediately permitting perfusion of the coronary arteries by the systemic intra arterial pressure (Fig 18A,B,C,D).

Resuscitation of the Heart: If the heart has not stopped contracting during the preceding period of momentary interruption of coronary flow, and if the aortic valve has been rendered competent, cardiac contractions should now become progressively stronger. If the heart action is weak it may be assisted by manual compression and the bypass rate may be increased to augment coronary perfusion. In the unhappy event that complete aortic competence should not have been achieved the left ventricle may show a tendency to become greatly (and possibly irreversibly) distended, and if this is so it will require prolonged assistance by manual compression.

Should ventricular fibrillation already be present or develop the administration of *lidocaine* intravenously (1 mg per kg) and electric countershock usually will restore an effective beat. Administration of lidocaine (100 mg in 500 cc.) by continuous drip may serve to prevent a recurrence of this untoward situation.

Calcium chloride or *gluconate* (1 Gm. intravenously), *digitalis* in one of its quick acting forms, and *epinephrine* (both as a single injection and as a drip) often have proved most valuable at this

juncture in strengthening a flagging heart *Norepinephrine* is useful in elevating the level of the arterial blood pressure and in maintaining it

Termination of the Bypass: As soon as the cardiac action becomes vigorous enough and the blood pressure becomes stabilized at an acceptable level, the rate of the perfusion will be reduced. If hypothermia has been utilized, rewarming will be started even before clamping the aortic incision by circulating warm water through the heat exchanger Perfusion, perhaps at a subtotal level, should be continued (from 1000 to 1500 cc per minute) until the body temperature has been elevated to a level sufficiently high to ensure continued spontaneous rewarming (usually at 33° C).

When all seems satisfactory the pump is stopped and the "arterial" and "venous" lines of the extracorporeal circuit are clamped Providing the blood pressure and cardiac action remain satisfactory preparations for decannulation are begun If not, resumption of the perfusion at a rate sufficient

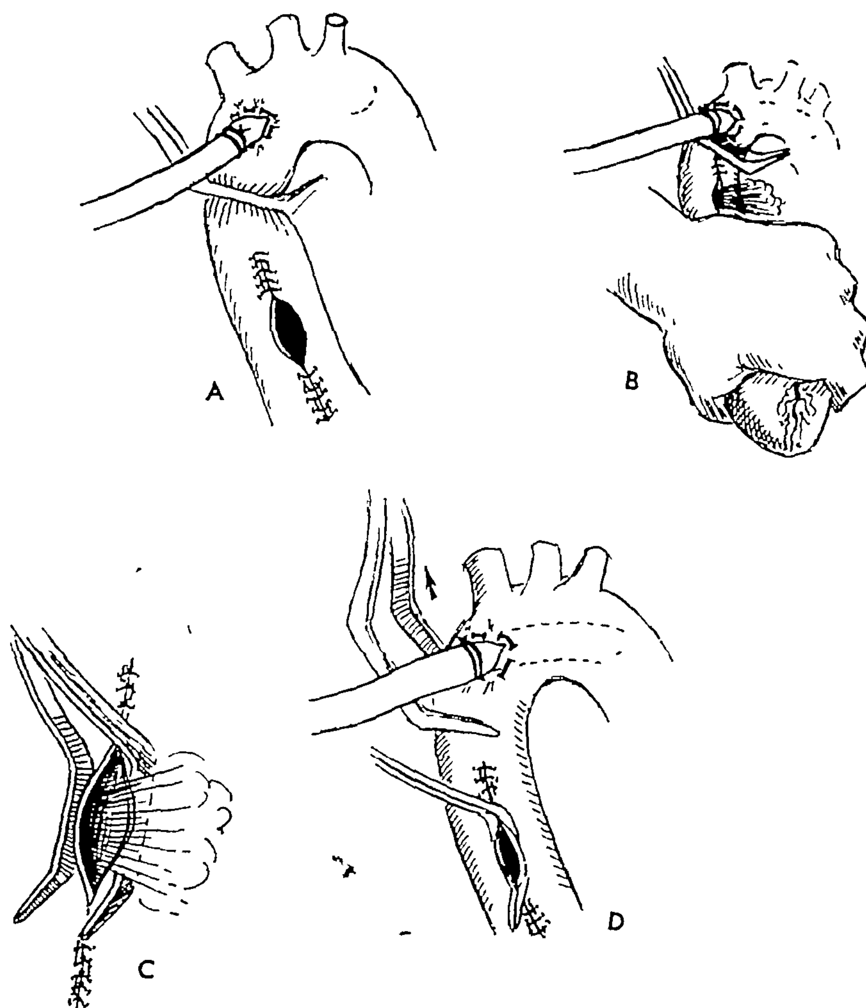


Fig 18 Restoration of circulatory integrity *A* The upper and lower thirds of the arterial incision are repaired with two layers of fine arterial silk The coronary catheters are removed *B* The pulmonary arterial tourniquet is released and the heart is compressed vigorously *C* When each compression expels a good volume of blood from the aortic incision a noncrushing clamp is applied to the residual aortic opening at the height of such a compression Thus all air is expressed from the heart and aorta *D* The aortic cross-clamp is removed immediately The remainder of the aortic wall is repaired with two rows of fine silk sutures

to restore and maintain myocardial vigor is in order. An attempt is made, thereafter gradually to "wean" the patient of such assistance. During this period closure of the remainder of the aortic incision may be carried out using one row of continuous mattress sutures, and one row of running sutures of fine silk (4-0 arterial).

Decannulation: The lips of the appendageal incision are picked up with fine hemostatic clamps, the sutures encircling the "venous" catheter are divided with a scalpel, and the appendage is clamped as this tube is withdrawn rapidly from the heart. The appendage is ligated proximal to the clamp with heavy braided silk (#2). The clamp is removed and the terminal incision is oversewn securely with fine arterial silk (4-0).

A 4-0 arterial silk suture is placed in the aortic wall at the cephalic extremity of the cannulation site, and another is affixed at its caudal extremity. The sutures encircling the catheter are divided and a dentate (Beck) clamp is applied to close the diminutive vascular incision as the catheter is withdrawn. The long ends of the silk sutures are used to repair the artery with one continuous mattress and one running suture line.

Restoration of Coagulability to the Blood — *Polybrene (Hexadimethrine Bromide)* is administered by intravenous drip in dosage of 1.5 to 2 mg. for each milligram of heparin the patient has received. The effective total dose of the latter is calculated as the sum of the initial body heparinizing dose, plus any subsequent such dose, plus one half of the total amount of heparin which was contained in all the blood added to the extracorporeal system. Twenty minutes after completion of the polybrene administration, the blood coagulation time is measured by the Lee White technic, and other hematologic studies are carried out. If a significant coagulation defect is recognized it is treated as indicated. Usually this consists of the intravenous administration of freshly drawn (citrated) whole blood, of fibrinogen, or of a suspension of preserved platelets.

Monitoring After Surgery: The saphenous and femoral arterial monitoring tubes may be kept open 24 to 48 hours by the slow continuous or frequent intermittent introduction of a very dilute solution of heparin sulfate in 5 per cent glucose. The respective venous and arterial pressure levels necessarily are obtained, therefore, only intermittently. The requirements for and the appropriate rate for administration of fluids or blood during the immediate postoperative period are regulated chiefly by these determinations.

Reconstruction of the Aortic Valve

Type I. Lesions. The aortic valve is inspected and the type of lesion and its pathophysiology is evaluated. In Type I pathology (normal cusps, dilated aortic root) a technic of "bicuspidization" by reduction of the number of aortic cusps to two has served admirably. This principle also was conceived independently by Garamella and associates[15] and by Creech,[16] although first accomplished by one of us (C.P.B.) on May 21, 1957,[6] with survival. The right atrium is dissected carefully away from the line of attachment of the noncoronary cusp. Then an ellipse of aortic wall (appropriate to the state of dilatation or size of the ascending aorta) is removed chiefly from the right lip of the aortic incision. The excised fragment should include nearly all of the noncoronary sinus of Valsalva, leaving however a small margin along the line

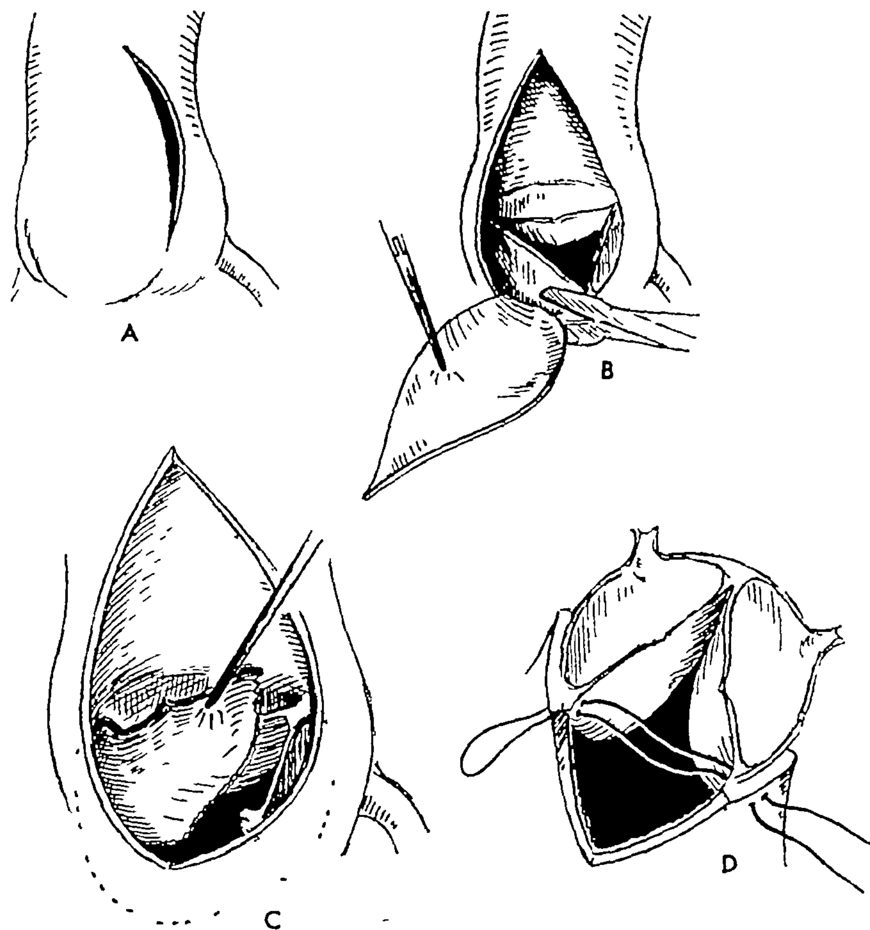


Fig 19 Bicuspidization of the aorta for Type I incompetence *A* Curved incision which has been made in the ventral aortic wall *B* Excision of an elliptical segment of the aortic wall including the noncoronary sinus of Valsalva *C* Excision of the noncoronary aortic cusp with preservation of the fibrous line of attachment to the arterial wall *D* Placement of traction sutures through the two commissures which are situated at either extremity of the by now excised noncoronary cusp

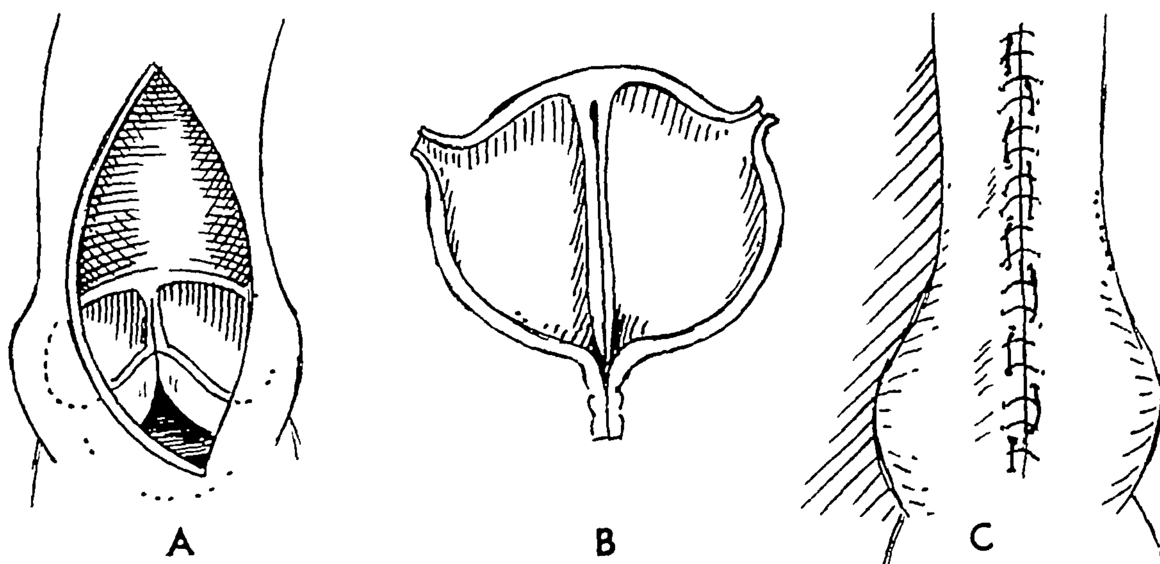


Fig 20 Closure of the created defects of the aortic wall and valve *A* Approximation of the dense fibrous condensations representing the attachment of the excised cusp *B* Top view of constructed bicuspid valve structure Note that the free margins of the remaining two cusps now lie in contiguity as parallel diameters of the reduced circular aperture *C* Note graceful (normal type) narrowing of the aortic caliber just beyond the level of the remaining sinuses of Valsalva This imposes an element of slight impedance to forward flow of the ejected blood Hence, there is an increase in lateral pressure which augments coronary perfusion

of cusp attachment. The noncoronary valve cusp is resected, leaving about a millimeter of its tissue attached to the aortic wall.

A fine but strong silk suture (two-zero) is passed through the two commissures which have bounded the now excised cusp and gentle traction is cusp Finally this suture is tied to the tightened and tied ends of the stitch in the commissures. This line of repair is reinforced by a second row of continuous running stitches (four zero-silk) (Figs. 20A B C and 21A,B,C)

Type II. Incompetence: Primary leaflet incompetence may be divided for



A

B

Fig. 21 Correctio of Type I aortic insufficiency by bicuspidization. A. Preoperative aortographic study showing 3 to 4+ aortic regurgitation and nearly aneurysmal dilatatio of the ascending aorta in a 42 year old white female who presented a history of previous lues. B. Postoperative aortographic study showing complete correction of valvular incompetence. Note the "tailoring of the formerly dilated ascending aorta.

made upon the ends to delineate the appropriate direction of suturing for the aortic repair (Fig 19A,B C,D) Then the lower portion of the aortic defect is closed by placement of one row of continuous silk mattress sutures (three zero) through the strong fibrous condensations, which represent the former lines of attachment of the noncoronary therapeutic purposes into fenestrations, prolapses, and retractions.

Fenestrations In this condition one or more perforations will be seen in one or more of the leaflets. Usually the cusps themselves will be found flexible and of approximately normal size and configuration although an element of shrinkage or prolapse may be asso-

ciated. In more complicated problems technics of an intermediate or combined type may be required.

In simple perforation a patch of prosthetic material or (preferably) a full thickness graft from the aortic wall is tailored to an appropriate size and shape and is laid within the concavity of the cusp over the perforation It then is fixed securely to the valve tissue with several mattress sutures of fine silk (3 or 4-0) applied at strategic points about the circumference of the defect Great care must be taken that the cusp does not become distorted by the sutures This leaves the patient with an effective tricuspid valve mechanism with only a slight functional reduction due to some loss of mobility of the repaired cusp. Restoration of complete valvular competence is the rule (Fig 22A,B,C,D,E).

As far as we know, this principle was applied for the first time on February 3, 1959 at the Hahnemann Hospital in Philadelphia

Prolapse In prolapse of one or more of the cusps it is essential, in order to obtain a high degree of correction of this type insufficiency, that at least one of the three cusps be relatively normal in size, shape, and flexibility, or capable of being rendered so. Whether it is one or two of the cusps that have prolapsed is not important since functional restoration is accomplished by su-

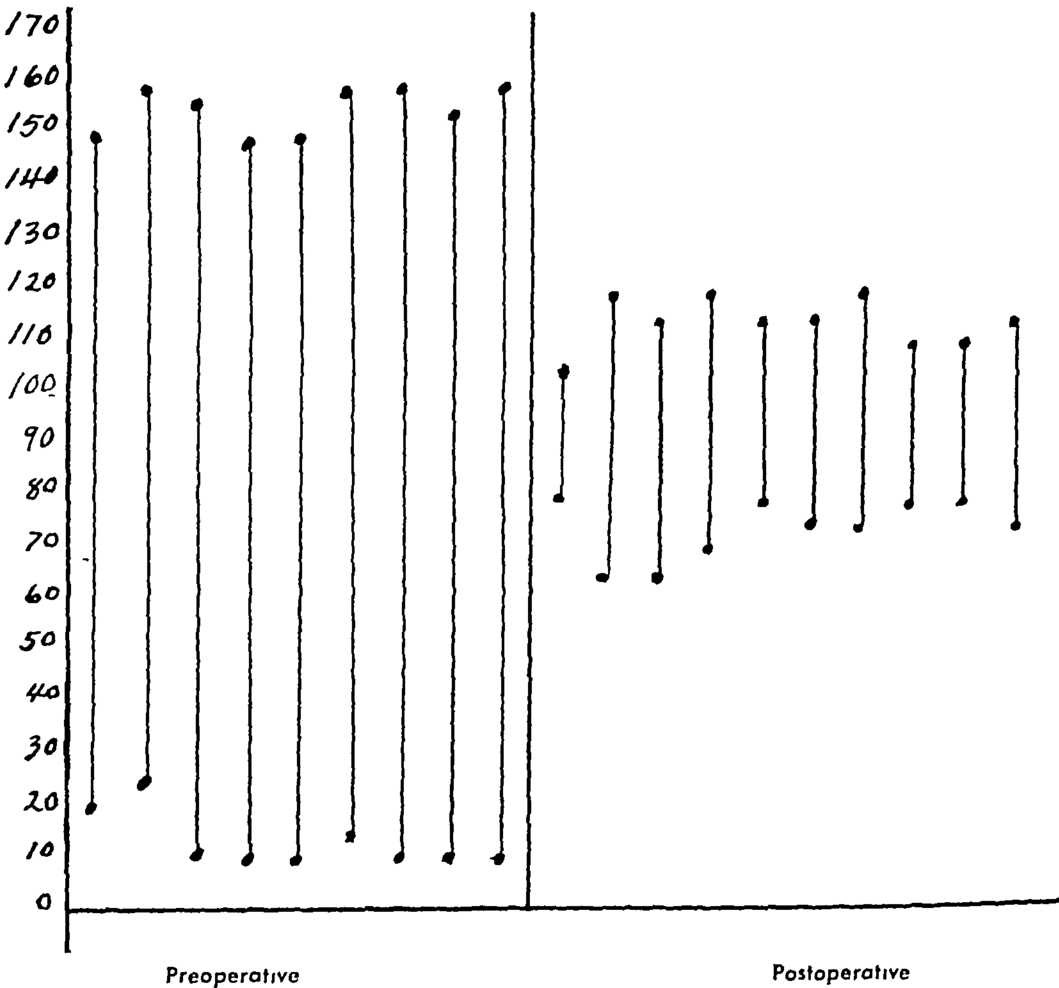
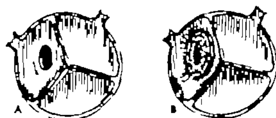


Fig 21C Pre- and postoperative blood pressure determinations

turing two cusps together to form a conjoint one. Thereafter while this conjoined valve element may exhibit appreciable limitation of mobility it will at least support itself by virtue of its attachment to two-thirds of the sponge, lest, under the stress of the incessant diastolic pounding, the sutures should tear out of the often very delicate valve tissue. It is important for a high grade of correction that the level at which the free edge of the conjoint



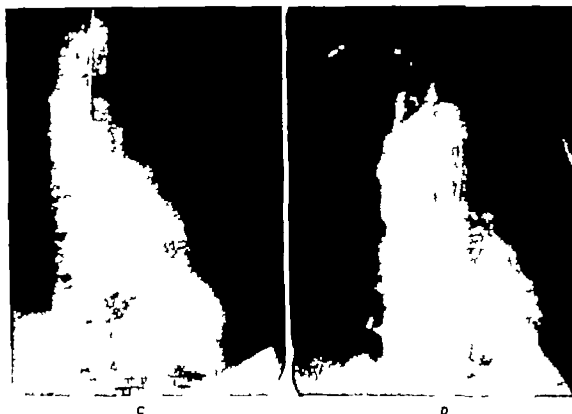


Fig. 22: Surgical correction of aortic incompetence due to perforation of a valve cusp. *A.* Perforated cusp in an otherwise normal valve. *B.* Fixation of a prosthetic patch or (preferably) a free segment of aortic wall within the concavity of the deficient cusp. *C.* 4-plus aortic insufficiency in a 36 year old white male patient demonstrated by aortography. *D.* Postoperative study showing 1 to 2 plus residual insufficiency. While the perforation was closed securely the additional valvular deformity (shrinkage of leaflets) was corrected only partially.

aortic circumference and will serve as a shelf against which the remaining individual cusp may abut during diastole (Fig 23A B C). It is preferable that the line of cusp suturing be buffered by the incorporation of strips of ivalon cusp comes to lie should not be above that of the free cusp (Figs. 24A B and 25A,B,C). Such a valve may be considered to have been converted into a bicuspid one of a somewhat different type, or if the conjoint cusp is rela

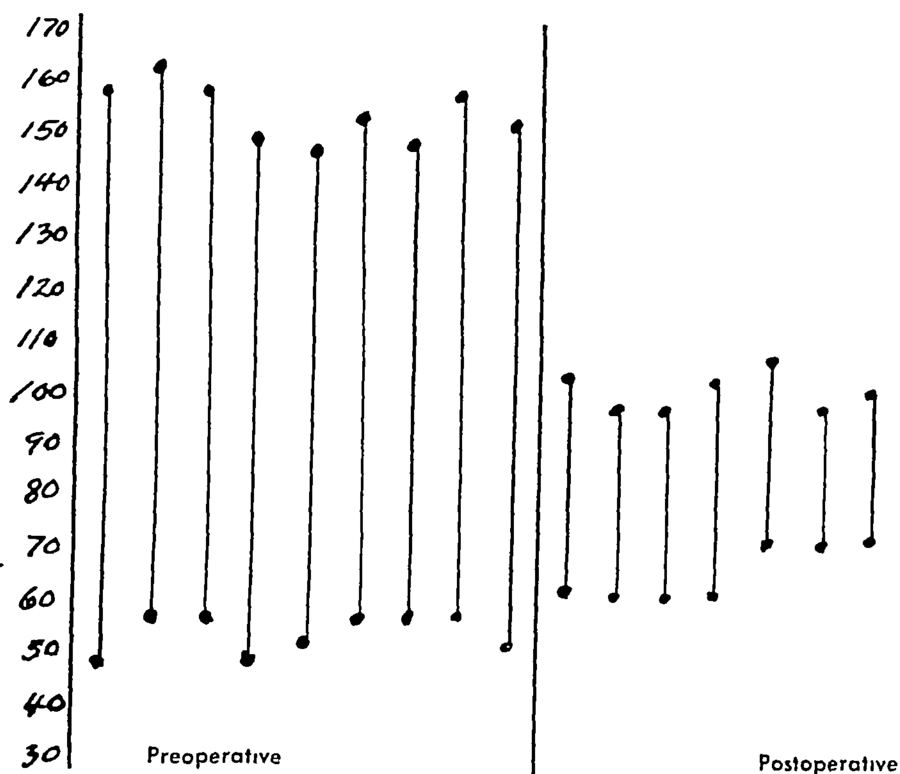


Fig 22E Blood pressure chart in same patient

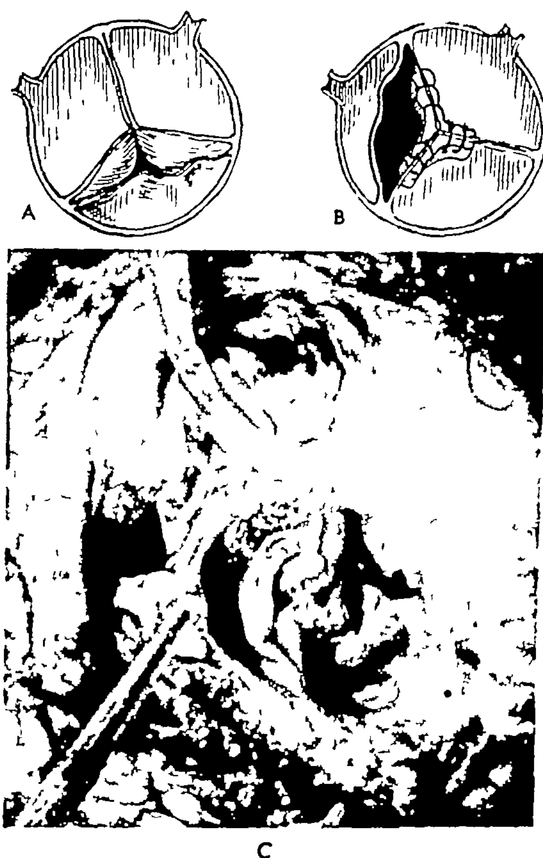


Fig 23 Prolapse of a single aortic valve cusp (Fig 3, p 118) corrected by suturing to one of its normal fellow cusps Two prolapsing cusps similarly may be united to form a single conjoint valvular shelf which then will be well supported by virtue of its attachment to two thirds of the aortic circumference A Preoperative appearance of the valve (from above) B Postoperative appearance of the valve (from above) Note the use of thin strips of ivalon sponge to b' the suture line C Autopsy specimen showing formerly incompetent aortic valve repaired suturing two cusps together (closed position)





A B

Fig. 24: Incompetence due to presentation of cusps at different levels within the aortic channel. A. Autopsy specimen showing an aortic valve with abundant leaflet substance but which had presented maximal incompetence because of malalignment and prolapse at the single fused commissure. The free margin of the noncoronary cusp presents at a lower level than the other two. B. Correction of this deformity in the autopsy room by dividing the fused commissure and resuturing it in such a way that the lower cusp now presents on the same level as the normal ones. The incompetence would have been overcome by this procedure had it been carried out during life.





A B

Fig. 25: Correction of aortic insufficiency in a 34 year old white female nurse who was found to have a prolapsing right coronary cusp. This cusp was sutured to the noncoronary leaflet thereby converting the valve into a bicuspid one. A. Preoperative aortogram (by suprasternal puncture) demonstrating 4-plus aortic regurgitation. B. Postoperative aortogram (by brachial arterial catheterization) demonstrating practically complete correction of the incompetence.

tively fixed or rigid, into an effective unicuspid one The first operation of this type was performed by C. W. Lillehei.[17]

Retractions—Shrinkage or retraction of one or more of the cusps in some instances is caused by rolling or retraction of the valve edges toward the respective sinuses of Valsalva, in others the deformity is due to actual shrinking of leaflet substance In the latter instance the valve tissue tends to be thickened and fibrosed As in cases of prolapse it is essential for restoration of valve competence that at least one cusp should be functionally effective or capable of being rendered so

Again the correction is accomplished by suturing together the remnants of two cusps, permitting the remaining less involved cusp by its independent movement to supply a unicuspid type of valve action. However, because of extreme valve retraction or shrinkage, approximation by direct suturing of the two more damaged cusps sometimes might eventuate in the creation of a tight shelf which comes to lie at a level significantly higher than that of the free edge of the remaining cusp. Hence, a new and perhaps equally severe type of valvular insufficiency may be imposed. Because of the actual or relative death of natural valve tissue

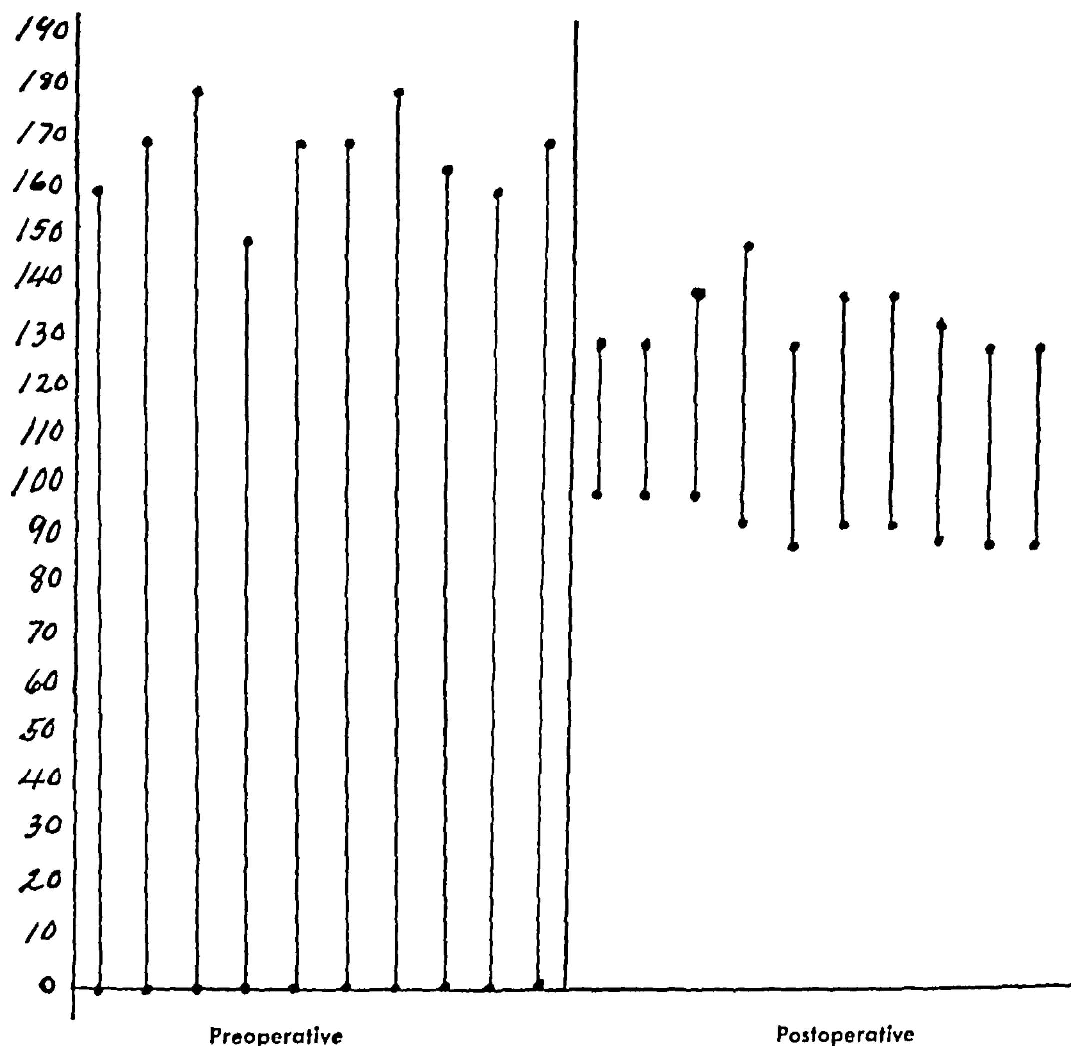


Fig 25C Pre- and postoperative blood pressure chart in same patient (Fig 25A B) showing favorable alteration in the circulatory dynamics (Bailey, C P, and Zimmerman, J: Derra's Handbuch Der Thoraxchirurgie, Springer-Verlag, 1959, p 970)

it sometimes becomes necessary to add substance either by grafting or by the use of a prosthetic.

In our practice it has seemed simplest merely to prepare a strong diaphragm from a "sandwich" of two layers of compressed ivalon which embrace a sheet of teflon netting. This "sandwich" is cut to appropriate size and shape and is sutured beneath the two cusps which are to be conjoined. The fibrous condensations of the aortic wall along the lines of leaflet origin are utilized for strong suture attachment. Then the overlying cusp tissue is merely affixed to this prosthetic diaphragm (Fig 26A,B,C,D). This procedure was carried out for the first time in a clinical case at the Flower and Fifth Avenue Hospitals, New York, February 26, 1960.

After creating a functional conjoint cusp or a rigid shelf of the type described, the remaining least involved cusp must be brought to a state of reasonable functional capacity. Should either "rolling" of the valve edge or fibrotic retraction prevent its proper functioning it may be deemed necessary to excise the cusp and to replace it entirely with a suitable autogenous graft (aortic wall tissue) or prosthetic material (Fig 27A,B,C,D). However in most instances it will be feasible for functional restoration merely to elongate the leaflet or to effectively "evert" it by suturing a crescentic fragment of the aortic wall as a free graft to the marginal zone and the bounding commissures (Fig 28A,B,C). Since the ascending aorta usually will be found dilated significantly a suitable amount of arterial tissue readily may be obtained from one of the lips of the aortic wound. This is facilitated by the presence of a curvilinear incision in the aortic wall (see Fig 16 (inset) and Fig 27B).

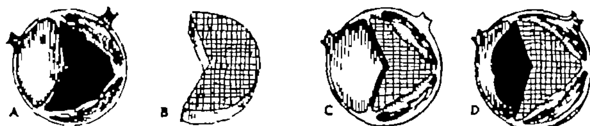


Fig 26. Method of overcoming aortic insufficiency when it is due to severe leaflet retraction. It is essential that at least one cusp remain functionally adequate or be capable of being rendered so at the time of operation. A. Severe retraction of the right coronary and the noncoronary cusps. The left coronary cusp (we hypothesize) is normal. B. A thick compressed prosthetic "sandwich" consisting of two layers of ivalon sponge with an intervening sheet of teflon mesh is "tailored" to provide a shelf with which the functional left coronary cusp can make contact during diastole. C. Prosthetic shelf sutured proximal to (below) the retracted cusps. The fibrous lines of cusp attachment are utilized to ensure firm suture fixation. D. Shrunken cusp remnants sutured to cephalad surface of shelf. Note divergence of functional left coronary cusp from fixed shelf during ejection, and convergence (Fig 26C) during diastole.

Type III Regurgitation ***(with Aortic Stenosis)*** Here a significant element of stenosis coexists with a serious grade of insufficiency. Commissural obliteration is the rule, and in most cases the cusps are severely fibrosed and exhibit a varying amount of calcific infiltration or encrustation (Fig 5).

Here it will be necessary before treating the incompetence at least to reestablish an adequate valvular passageway. If possible, an appreciable

Fig 27: Method of providing a functional third cusp in the presence of an immobile conjoint cusp or prosthetic shelf *A* Total excision of the remaining single cusp may be required. *B* The excised leaflet may be replaced by a prosthetic cusp or by a crescent shaped free graft obtained from the aortic wall *C* Placement of graft or prosthetic cusp, top view *D* Side view of same

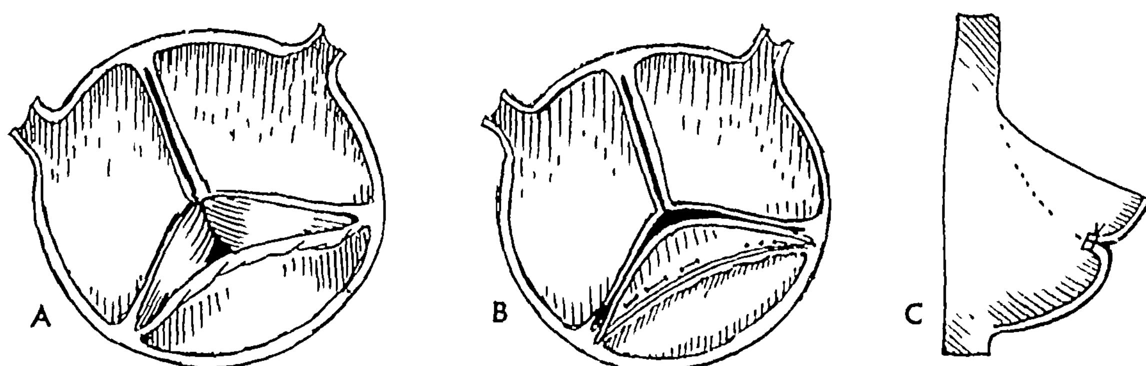


Fig 28: Elongation of a shortened or everted cusp by suturing a free aortic wall graft to the rolled margin *A* Rolled margin and retraction of noncoronary cusp *B* Noncoronary cusp elongated by addition of crescentic graft to free margin *C* Sectional view of the same

degree of cusp flexibility also should be restored. These accomplishments will demand the exhibition of the technics of valvular "sculpturing" previously described.[18, 19] In some cases the improved flexibility of the valve components will so contribute to valve competence that the insufficiency will be overcome completely or nearly so. In other instances because of extreme pathologic hardening or dearth of sufficient valvular tissue competence cannot be reestablished in this manner.

Reverting to the expressed principle that at least one functional cusp must be provided the surgeon usually will utilize the principles of treatment applicable for Type II lesions especially those shown in Figures 26, 27 and 28.

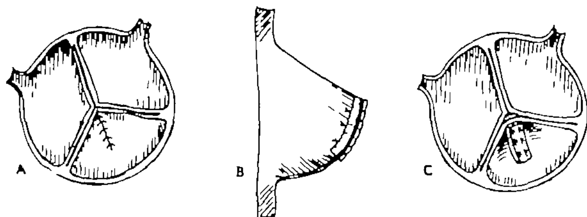


Fig 29 Repair of a divided leaflet. A "sandwich" technic provides the strongest possible reinforcement of a lacerated cusp. A. The divided edges are sutured together primarily whenever this is possible. B. A thin layer of compressed Ivalon sponge is placed over the convex surface of the cusp. A second layer of compressed Ivalon sponge reinforced with teflon netting is applied within the concavity of the cusp. C. Through and through mattress sutures of arterial silk establish firm attachment and reinforcement.

Type IV Regurgitation (*Dissolution of Leaflet Continuity*): Since the possible variations in the anatomic lesion are infinite the treatment in any particular case must be individualized. When the defect amounts to a perforation of a leaflet a "liner" of prosthetic material or autogenous arterial wall may be placed within the concavity of the affected cusp to cover the opening as illustrated in Figure 22A,B. When the valve margin has become separated continuity may be restored by "patching" with prosthetic material. Usually it is best to use a "sandwich" technic applying a thin slab of plastic fabric or substance to both the convex and the concave aspects of the injured leaflet. Through and through sutures then will suffice to hold the valvular remnants until healing has taken place (Fig 29A, B,C).

Should the aortic root be dilated significantly and should the lesion be limited to the noncoronary cusp this may be excised along with the noncoronary sinus of Valsalva. Then repair of the aortic wall will convert the valve into a truly bicuspid one as in the correction of Type I lesions (Fig 20A,B C).

Obviously these suggestions for the repair of Type IV lesions may be inapplicable if more than one cusp is damaged and in those cases in which the valve elements are significantly inflexible. In them, technics similar to those illustrated in Figures 26 and 27 may be more suitable.

Summary and Conclusions

Aortic insufficiency amounts essentially to an aortico-left ventricular fistula, and its clinical seriousness depends largely upon the magnitude of the leak The types of lesion which can bring about incompetence of the aortic valve are many For each individual variation it now is possible to postulate a suitable corrective technic For the majority of cases conversion of the tricuspid valve mechanism into a bicuspid one has provided the blue-print for success In some instances the final result is a unicuspid valve, the single functional cusp abutting against a rigid shelf formed from the remaining two cusps with or without the additional utilization of prosthetic materials When no suitable functional leaflet exists or can be constructed from the valvular remnants a prosthetic cusp or one fashioned from available excessive aortic wall substance may be used after excision of the natural cusp structure

All of these procedures for valvular reconstruction are dependent for clinical success upon technics which recently have been devised to support the body circulation and the myocardial vigor during operation The body circulatory requirements may be provided for by one of the accepted perfusion systems, perhaps with the aid of hypothermia Cannulation of the coronary arteries and perfusion with arterialized blood during surgery permits maintenance of such cardiac tone that no difficulty is experienced in resuscitating the heart at the conclusion of the procedure

By shrewd utilization of these principles it is possible, when they have not progressed to the final stages, to operate on patients with aortic insufficiency with an acceptable surgical risk (10 to 15 per cent), and with every prospect that complete or near-complete valvular competence will be achieved Any unavoidable resultant reduction in the size of the valve orifice may be construed as a small price to be paid for correction of the aortic regurgitation

References

1 Bailey, C P, et al Attempts at Surgical Correction of Rheumatic Aortic Regurgitation Presented at Ann Meet. Am Heart Assoc, Cleveland, April, 1952
2 Bailey, C P., Glover, R P, O'Neill, J T E, and Redondo-Ramirez, H J Thoracic Surg., 20 516 (Oct.) 1950
3 Hufnagel, C A Bull Georgetown Univ M Cent, 4 128 (Feb-Mar) 1951
4 Hufnagel, C A, and Harvey, W P Ibid, 6 60 (Feb) 1953
5 Bailey, C P, and Zimmerman, J Aortic Regurgitation Encyclo Thoracic Surg, Springer-Verlag, Heidelberg, Germany, 1959, p 948
6 Bailey, C P., and Zimmerman, J Am J Cardiol., 3 6 (Jan) 1959
7 Burch, G E Am Heart J, 50 352 (Sept) 1955
8 Beuren, A., Sparks, C, and Bing, R J Am J Cardiol, 1 103, 1953
9 Blanco, G, et al Ibid, 2 302 (Sept) 1958
10 Zimmerman, J J Albert Einstein Med Center, 7 77 (Apr) 1959
11 Fitch, E A and Bailey, C P J Thoracic Surg, 37 660 (May) 1959
12 Nunez, L E. and Bailey, C P Ibid, Vol 37 707 (June) 1959
13 Kay, E B, Head, L R., and Nogueira, C J A M.A, 168 1767 (Nov 29) 1958
14 Bailey, C P., and Zimmerman, J Am J Cardiol, 3 13 (Jan) 1959
15 Garamella, J J, et al New Concept in Surg Treat of Aortic Stenosis Meet Minn Surg Soc, Nov 1, 1957, Minneapolis
16 Creech, O, Jr Bull Tulane M Fac., 17 73 (Feb) 1958
17 Lillehei, C W, Gott, V L, DeWall, R A, and Varco, R L J Thoracic Surg, 35 154 (Feb) 1958
18 Darra, E Encyclo Thoracic Surg, Springer-Verlag, Heidelberg, Germany, 1959, p 971
19 Gould, S E Pathology of the Heart. Charles C Thomas, Springfield, 1960, 2nd Ed

SURGERY FOR MITRAL STENOSIS

Dryden P Morse, M.D , Jacob Zimmerman F R.C.S., and Charles P Bailey M D

Mitral stenosis is the most commonly encountered lesion of the heart for which definitive surgery presently is available. Essentially this lesion is but a mechanical obstruction of the left atrioventricular valve. Nearly invariably it is the end state resulting from previous damage to the heart by the rheumatic process.

Because mitral stenosis is basically a mechanical lesion, it logically might be expected to lend itself well to correction by surgery Nevertheless, it is only during the past eleven years that surgical relief of this lesion has been successfully accomplished. And during these few years there has been continuing change and improvement both in concepts as to the objectives of surgical alleviation of the lesion and in the surgical technics available for its correction. The surgical objective at the beginning of this work was an elongation of the valve "slit"[1] (Fig 1 A and B) This generally sufficed to relieve symptoms without, however removing the obstruction at the valve completely This separation of one or both commissures was accomplished either by quick digital dilatation or by rather poorly controlled instrumental enlargement of the valve orifice.

More recent efforts however have been directed toward the complete alleviation of the obstruction and toward the reconstruction of a fully functional valve. This requires the utilization of a much more elaborate technic in order to create a unicuspid flap valve by mobilization of a new "septal leaflet" (Fig 2 A and B)



Fig. 1 Diagrammatic representation of presumed accomplishments of original "commissurotomy" operation for mitral stenosis. A Closed valve B Opened valve (O'Neill, T J E, et al J. Int Coll. Surg., 13:359 (Apr.) 1950.)

An understanding of the surgical concepts and technics which are fundamental to the modern procedure for the correction of mitral stenosis requires a reevaluation of all the pertinent anatomic, pathologic, and pathophysiologic considerations.

Anatomy

Anatomic Aspects of Particular Significance: The left ventricular myocard

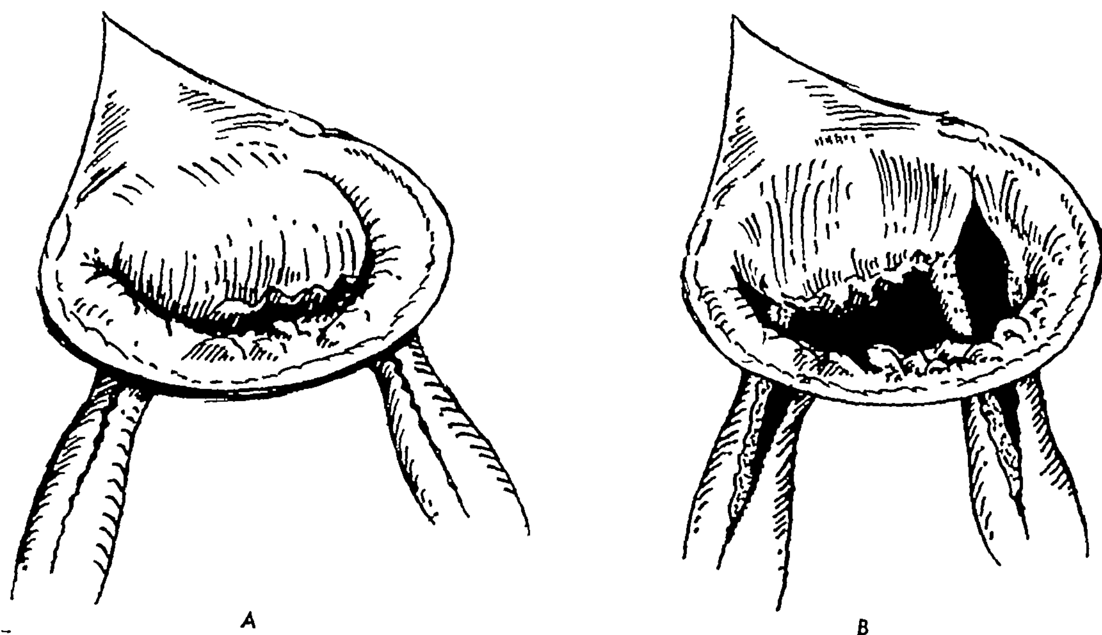


Fig 2 Neostrophingic mobilization of the stenotic mitral valve Note that a basally flexible tongue-like flap has been created from the longer or septal portion of the sleeve like structure This provides a unicuspid type of valve action The irreparably damaged mural portion of the sleeve acts merely as a rigid shelf against which the mobilized flap can abut to provide competence Chordopapillary agglutination is relieved by longitudinal splitting of the "fusion mass" This may be accomplished either by an open or a closed technic. A Mobilized valve in closed position B Mobilized valve in open position

ium can be visualized as a hollow muscular cone The single combined (aortic and mitral) orifice at the base of the cone allows ingress of blood across the mitral valve as well as egress through the aortic valve The "annulus" of the mitral valve is a horseshoe-shaped fibrous thickening which is disposed along the posterior aspect of the rim of the ventricular cone The fibrous horseshoe joins at its free ends with two dense bodies, the right and left fibrous trigones, which are situated at opposite sides of the ventricular orifice and extend about half-way around its circumference (Fig 3). As an anatomic structure the annulus is, as mentioned, deficient anteriorly The remaining anterior aspect of the ventricular orifice is occupied by the root of the aorta which is anchored to the left ventricular myocardium by the three low points of its valvular attachment To complete the *mitral* orifice, a fibrous curtain arises from the lower edge of the aorta, be-

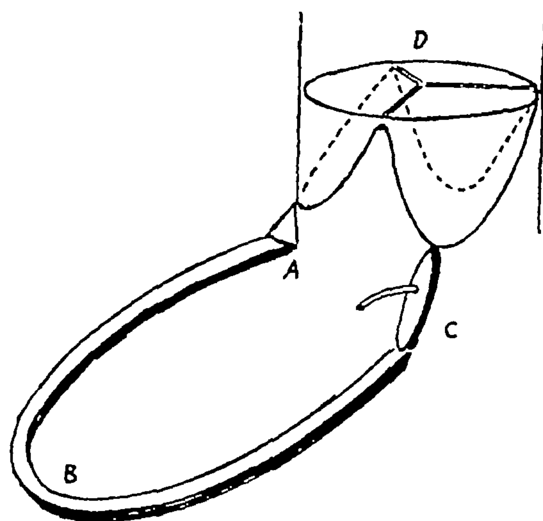


Fig 3 Schematic representation of mitral annulus fibrosus as seen from the atrial aspect (After a dissection by Zimmerman) Note that the "ring" is a horseshoe shaped structure while the aortic and pulmonary valves have no annuli unless the zig-zag lines of fibrous condensation which mark the sites of attachment of the respective semilunar cusps be so considered A Junction of "annulus" with the left fibrous trigone and the low point of the left coronary cusp B Mural portion of "annulus" C Junction of "annulus" with the right fibrous trigone and the "low point" of the noncoronary cusp

tween the attachment of the left and the noncoronary cusps, spans the distance between the two trigonal "points" and is continuous with the septal, or at times called aortic, "leaflet" of the mitral valve (Fig 4)



Fig. 4: Aortic and mitral (stenotic) valves exposed by wide incision of the left ventricle Note chordopapillary fusion which is both intrinsic and extrinsic (to the ventricular aspect of the mitral valve) The longer or septal side of the valve sleeve may be seen to arise from the subaortic "curtain" outlined by: A. The "low point" of the right coronary cusp. B The commissure between them. C The "low point" of the noncoronary cusp.

The Valve The mitral valve which classically is described as a bicuspid or two-leaflet structure is in fact a continuous sleeve or ribbon of membranous tissue of irregular width. The classically described "leaflets" are arbitrarily designated segments of this continuous although irregular sleeve of tissue. However the "septal leaflet," while anatomically and histologically inseparable from the remainder of the valve sleeve, is somewhat different functionally in that it is the portion which becomes "bulged inward in convex fashion as the "ribbon" becomes "enfolded" during ventricular systole (Fig 5) The entire valve may be visualized as a membranous cylinder with an obliquely truncated base (Fig 6 A and B) The longer side then constitutes the portion which generally is designated as the septal leaflet.

Subvalvular Supporting Structures The chordae tendineae of the first order are attached along the free edge of the valve sleeve. Secondary and tertiary



Fig. 5: Normal mitral valve in closed position as seen from the atrial aspect Note the line of closure. Note also that the struct re is a continuous ribbon or sleeve of membranous tissue with an irregular and delicately convoluted edge No discrete cusps can be discerned (Bailey C. P.: Surgery of the Heart Lea and Febiger Philadelphia, 1955 Ed 1.)

chordae arise from the ventricular surface of the valve cylinder (Fig 7) The primary chordae end in two papillary muscles which underlie the two extremities of the normally arcuate line of valve closure.

The mitral valve does not close by the coaptation of opposing "leaflets but by the centripetal folding together of the complete valve sleeve toward the center of the orifice. The resulting valve edge contact, as viewed from the atrial aspect, is a slightly irregular ar

cuate line (Figs 5 and 6 B), the extremities of which, if projected, would extend directly to the points of attachment of the annulus fibrosus with the respective fibrous trigones These are

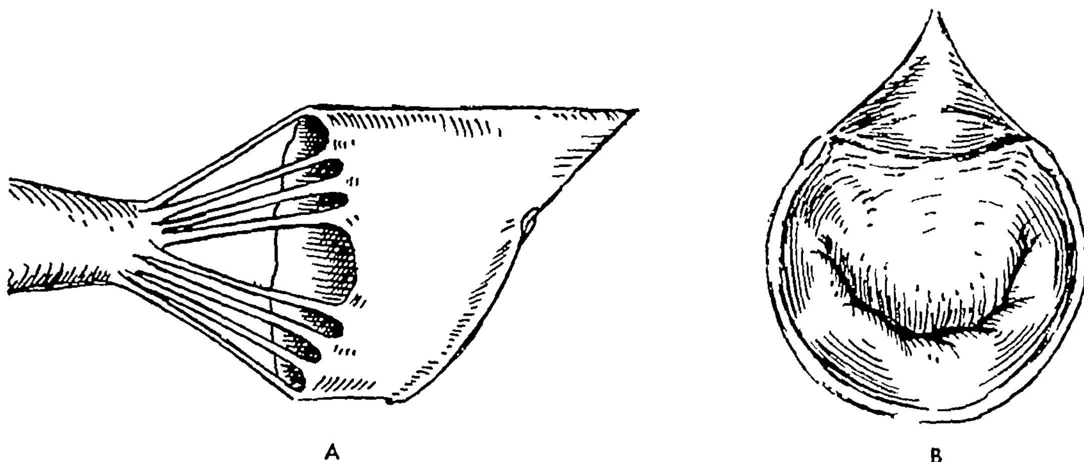


Fig 6 A Diagrammatic illustration of the normal mitral valve cylinder during diastole B During ventricular systole the valve closes along an arcuate line (Fig 5) in response to the respective amounts of "hydraulic press" which are accepted in proportion to their area by the unequally wide portions of the valve sleeve

the so-called surgical "target" points At the beginning of diastole the delicately convoluted edge of the valve sleeve moves effortlessly apart and



Fig 7 Ventricular view of the normal mitral valve showing the sleeve like character of the structure The free margins and ventricular aspect of the valve are supported by chordae of three orders They are notably deficient over the longer septal or aortic aspect of the "valve cylinder" (Bailey, C P, et al New York State J Med, 56 649 (Mar 1) 1956)

ahead of the atrial blood as it flows into the ventricular cavity The arcuate line of valve closure during systole can be seen to be but a logical response to the hydrodynamic alterations during systole since the greater surface area of the wider "septal portion" necessarily is subjected to a greater total force than is the narrower (mural) portion of the valve sleeve

Pathology

While the rheumatic process may involve the entire heart including the myocardium and the pericardial surfaces, maximal inflammation usually is expended upon the endocardial surfaces The reaction to inflammation is intensified upon the free edge and over the contact surfaces of the valve because of the incessant trauma incident to opening and closing Not only these surfaces but also those of the chordae tendinae and the papillary muscles may take part in the inflammation and become enveloped with *fibrinous exudate* (Fig 8A,B,C) The organization and fibrosis of the covering exudate and the inflamed surfaces which follows during

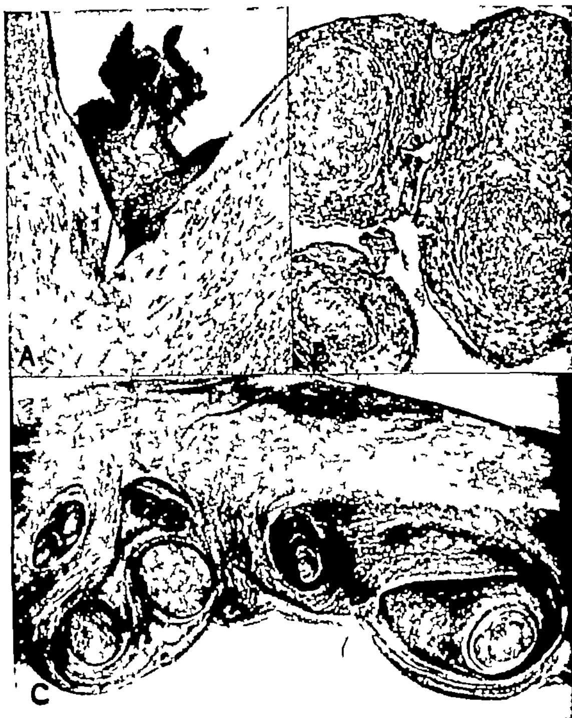


Fig 8. Changes which take place upon the surfaces of the valve mechanism. A. Not only does fibrinous exudate tend to envelope the entire valve mechanism (and especially its atrial surface) but it becomes partially dislodged and "heaped-up" at the sites of normal folding of the valve ribbon. Subsequent organization of this exudate converts it into fibrous connective tissue which coats the structure rendering it thicker and less pliable. Cross-adherence of the ribbon at the extremities of the arcuate line of closure may produce pathologic angles or "commissures." B. Pathologic adherence of contiguous chordae tendineae due to organization of the exudate which has enveloped and agglutinated them in "clumps." C. Cross-section view of rheumatic mitral valve ribbon showing pathologic adherence and partial incorporation of chordae of the second order within the thickened ventricular aspect of the valve. (Magarey F R, Brit. Med. J., 1 856 1957.)

the healing phase result in shortening and sometimes in crossagglutination of these supporting structures, and in thickening and purse-stringing of the normally thin, delicate edge of the valve. Thus, the valve "cylinder" becomes converted into a valve "cone," the base of which, of course, remains obliquely truncated (Fig 9A,B,C,) At the extremities of the shortened (purse-stringed) arc of closure the thickened valve margins tend to form relatively rigid "commissures" Not infrequently but certainly not constantly a certain amount of crossfusion of the opposed valve margins will take place beginning at the "commissures" and progressing in a centripetal manner (Fig 10A,B,C,D,E,F,G)

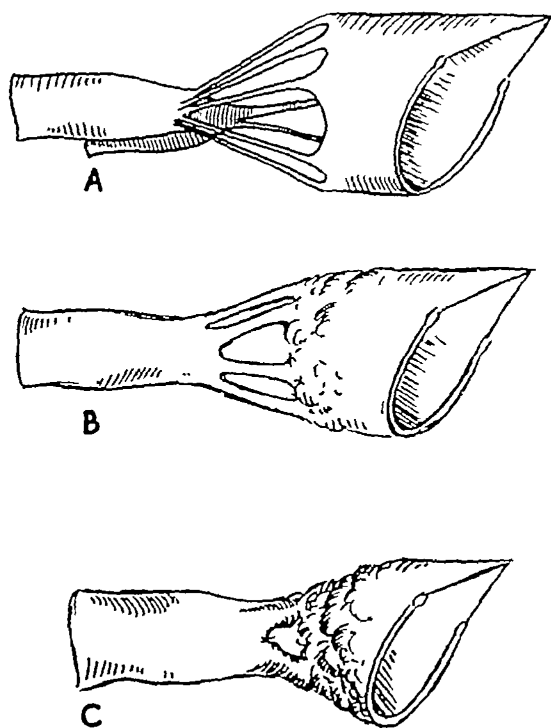


Fig 9 Development of Mitral Stenosis A Normal valve cylinder B Beginning organization of vegetations and enveloping exudate deposited upon the distal of marginal (inflamed) portion of the valve cylinder The width of the zone of valve inflammation averages about 1 cm C This process in addition to the healing of the intrinsic lesions within the cusps (foci of fibrinoid destruction) leads to "purse stringing" of the distal portion of the "cylinder" converting it to a "valve-cone" This cone retains the original obliquely "truncated" base The proximal or basal half of the longer portion or "side" of the cone having been spared by the inflammatory process remains thin and flexible It is this portion which provides the "hinge" of the tongue like flap which is provided by "neo strophingic mobilization"

Pathophysiology

As it progresses the narrowing of the mitral valve opening tends to impede blood flow and to cause back pressure which frequently produces considerable enlargement of the left atrium In turn, the elevated left atrial pressure is transmitted backward through the valveless pulmonary veins causing an increase in the pulmonary capillary and later in the pulmonary arterial pressure At first, these phenomena manifest themselves only upon exertion The amount of physical effort required to produce them becomes progressively less as the opening of the valve becomes smaller until finally the left atrial pressure is elevated even at rest, rising to still higher levels on exertion

The physiologic effects of mitral stenosis may be reflected indirectly in the form of decompensation of the right ventricle with peripheral edema (because of the burden imposed by the pulmonary arterial hypertension), or they may be evidenced more directly in the results of increased pulmonary capillary pressure in the form of acute pulmonary edema, hemoptysis, or chronic nonproductive cough. The distention of the left atrium tends to bring about atrial fibrillation and this is seen in more than 50 per cent of these patients at the time of first consultation with the surgeon

Clinical Findings in Mitral Stenosis

Symptoms: A moderate degree of mitral stenosis may not produce symptoms for years A murmur may be picked up during routine physical ex-

amination. Symptoms not infrequently are precipitated by pregnancy or some other condition which entails a considerable increase in the cardiac output (obesity, overwork, etc.) The most most prominent and common symptom in mitral stenosis. In most cases, it comes on insidiously over the years. At first it acts merely as a brake to the more extreme types of exertion but

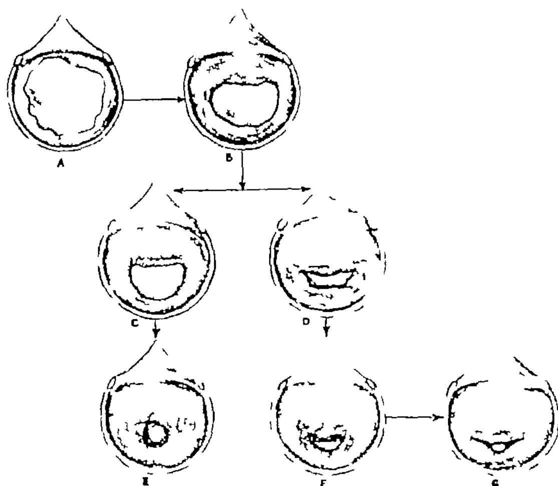


Fig. 10: Variations in the course of development of mitral stenosis as seen from the atrial aspect. A. Representation of normal mitral valve showing thin convoluted free margins. B. Organization of intrinsic lesions (areas of fibrinoid necrosis) and of externally deposited exudate covering the distal "zone of inflammation" leads to more or less symmetrical "purse-stringing" of the valve-cone apex. C. Further narrowing and hardening of the "zone of inflammation" leading to significant valvular obstruction. D. Continuation of this process converts the original "valve cylinder" to a valve-cone with a very tiny apical orifice but *without any suggestion of commissure formation!* E. On the other hand "freezing" of the indurated valve of stage C in a position of partial closure lends itself to commissure formation. F. Crossfusion of the hardened valve margins by organization of heaped up exudate accumulated at the commissures (Fig. 8A) rapidly reduces the diminished valve area. G. Final classical pin-point mitral stenosis with well-marked commissures.

common symptoms are shortness of breath and fatigue; others are cough, palpitation, weakness, and pain in the chest.

Shortness of breath on exertion is the later becomes sufficiently bothersome to restrict the patient to a sedentery existence. Not uncommonly the patient restricts herself unconsciously and therefore is not aware of the reduced

reserve until some unusual physical exertion is undertaken The progression of the mitral stenosis and the increase in the left atrial back pressure occasionally may be associated with shortness of breath at night which may awaken the patient (paroxysmal nocturnal dyspnea) In the later stages of the disease, orthopnea becomes a prominent symptom

The *cough* in mitral stenosis is due to congestion of the lungs and bronchial mucosa and sometimes is the patient's most annoying symptom The pulmonary hypertension may be accompanied by hemoptysis or frank pulmonary infarction, especially in the later stages of the disease

Palpitation represents either the occurrence of premature atrial beats or bouts of frank atrial fibrillation Atrial fibrillation tends at first to be episodic but with the progress of time becomes permanent in a high proportion of cases

Chest pain and a sense of tightness in the chest usually is attributed to the pulmonary hypertension, although the reduced cardiac output which occurs late in the disease may also cause a certain amount of functional reduction in the coronary blood flow.

In rare cases, *hoarseness* may develop This appears to be the result of compression of the left recurrent laryngeal nerve between the enlarged main or left pulmonary artery and the under surface of the aortic arch *Arterial embolization* is said to occur in approximately one-third of patients with pure mitral stenosis Usually, this is the result of dislodgement of thrombotic material from the left atrium or atrial appendage Embolization is rare in the absence of atrial fibrillation Common sites for embolic localization are the cerebral, abdominal, and limb arteries The symptoms are determined by the location and the size of the artery involved.

Right heart failure causes swelling of the ankles, a general gain in weight and hepatic and peripheral venous distention The effects of tricuspid insufficiency are similar, but with the addition of pulsating neck veins and disproportionately prominent hepatomegaly and ascites

Usual Course: After one or more typical or atypical episodes of rheumatic fever during childhood or adolescence, the patient becomes and remains "well" for many years She then develops shortness of breath on exertion and occasionally may note palpitation of the heart After a certain period of comparative symptomatic stability, the symptoms rapidly become more severe and disabling and right heart failure impends. Systemic arterial embolization may punctuate the course at any time

Mitral stenosis differs from other valvular lesions of the heart by its characteristically long history of dyspnea and orthopnea and its rather ready, although often temporary, response to rest and other medical therapeusis. In mitral insufficiency, by contrast, fatigue is more marked than dyspnea, and orthopnea seldom occurs until the latest stages have been reached

Physical Signs: The irregular pulse of atrial fibrillation is found in approximately one-half of the patients. There is a midlate diastolic, or holodiastolic murmur at the apex which commonly is described as rumbling. It is heard best towards the left axilla when the patient assumes the lateral recumbent position on his left side Often a diastolic thrill which is the palpatory expression of the same vibration will be detected There may be a loud "slapping" first

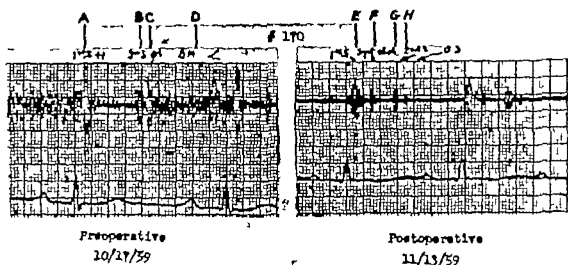


Fig. 11: Fairly typical pre- and postoperative phonocardiographic findings in mitral stenosis corrected by the neostrophingic method A and E—first heart sounds. B and G—second heart sounds. C (O*)—opening snap. D (DM)—diastolic murmur F—systolic "click."

sound at the apex with a 'presystolic accentuation of the diastolic murmur and in some cases, an "opening snap of the fibrotic mitral valve (Fig 11) Pulmonary hypertension is manifested by increased loudness of the second sound in the "pulmonary area. Often the second sound is split.

X Ray Features X ray studies and fluoroscopy demonstrate the left atrial enlargement by a widening of the waist" of the heart in the postero-anterior view and by a posterior displacement of the (barium filled) esophagus in the right anterior oblique and lateral views (Fig 12 A and B) Pulmo



Fig. 12 Fairly typical radiographic findings in mitral stenosis. A. Posteroanterior radiograph. Note the widening of the "waist of heart" and the prominence of the pulmonary vascular markings. B. Lateral view showing marked posterior displacement of the barium filled esophagus by the dilated left atrium.

nary hypertension may be connoted by increased prominence of the pulmonary arteries, and right ventricular enlargement by encroachment of the heart shadow upon the retrosternal air shadow in the lateral view. In late cases with long standing pulmonary hypertension, the peripheral lung fields may show the "miliary" nodules of chronic pulmonary congestion and hemosiderosis.

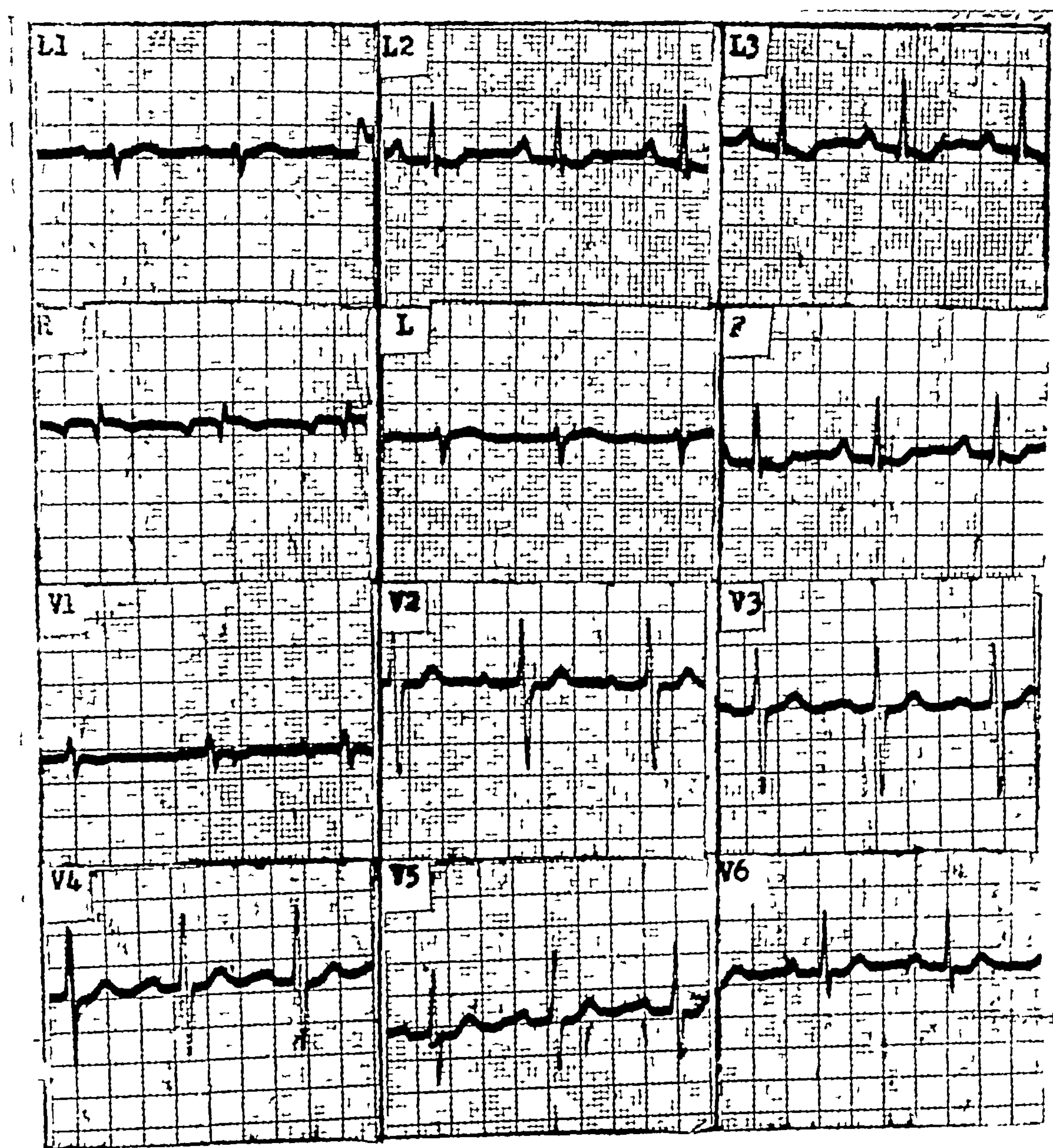


Fig 13 Electrocardiographic studies on patient from whom the phonocardiograms on Fig 11 were obtained Note prolongation of the P-R interval and CO 24 sec and prominence of the P wave Arrhythmias such as atrial fibrillation are very common in mitral stenosis

Electrocardiographic Changes: Certain distinctive features may be noted in the electrocardiogram in mitral stenosis. First, the P waves may be high, peaked, or notched The large area beneath the P wave is said to indicate a large and forcibly contracting atrium Second, right axis deviation and right ventricular hypertrophy of variable degree may be present Third, ex-

amination of the chest leads shows that the right ventricle forms the major portion of the anterior part of the heart since right ventricular enlargement is ever present. Disturbances of the rhythm and rate and prolongation of the P-R interval beyond 0.20 seconds are not uncommon (Fig. 13).

Diagnosis

The diagnosis usually may be made on clinical grounds alone. However, when multivalvular involvement is present or when the patient has been operated upon previously for mitral stenosis and there is some question as to whether *restenosis* has occurred, the diagnosis can be established best by the actual physical measurement of the pressure gradient across the valve by "left heart" catheterization. These data can be correlated with a determination of the flow across the valve to calculate the valve area. Any elevation in the left atrial pressure is reflected in an elevation of the "wedge" pressure (pulmonary venous capillary) as determined during right heart catheterization.

Frequently a question exists as to the presence of mitral insufficiency in a patient who has a systolic murmur at the apex but who otherwise presents evidences of mitral stenosis.

A systolic murmur may be caused by tricuspid insufficiency and this, also, may be heard best at the cardiac apex. Often such a murmur will be associated with unusually severe hepatomegaly and pulsating or distended neck veins. The murmur of tricuspid regurgitation tends to vary in intensity with deep inspiration and expiration more directly than do murmurs due to *mitral insufficiency*. Furthermore, some systolic tricuspid murmurs which are heard while the right heart is in a state of decompensation will diminish or even disappear after a few days of bed rest, digitalis, and diuretic therapy. On the other hand, the murmur of mitral insufficiency, especially in the older patient with a fibrotic or fibrocalcific valve, will not change much with therapy of this nature. The diagnosis in any of these cases may be resolved by injection of radio-opaque contrast material into the appropriate ventricle. This will demonstrate the competence or incompetence of the respective atrioventricular valve. The amount of regurgitation may be quantitated roughly by an estimation of the amount of contrast medium which enters the atrium during systole (Fig. 14 A and B).

Indications for Surgery

The indications for surgery in mitral stenosis have changed progressively as the operative technics have improved. At first, because of the relatively high risk of the operation, it was felt that intervention should be reserved for the severely symptomatic patient. It was felt also that it should be withheld from the patient who was in a state of intractable heart failure with advanced cardiac pathology. More recent improvements and innovations in the surgical technic, however, have so lowered the operative risk that both the "early" and the late stages of mitral stenosis now may be considered operable. It may safely be postulated that in mitral stenosis the pathologic state and the degree of physiologic obstruction always are "later than you think!" Present day criteria for surgery consist essentially of the demonstration either clinically or by cardiac catheterization of a significant degree of obstruction to the flow of blood through the

mitral valve, in the absence of any appropriate contraindication. The possibilities for salvage and for marked clinical improvement in the status of far advanced patients are much greater than is generally appreciated

Special Surgical Considerations

Restenosis: The indication for surgery in a patient who has had an operation for mitral stenosis, who has been improved or clinically well for a few years, and who then has had recurrence of his original symptomatology, is the demonstration of a significant gradient across the mitral valve by left heart catheterization (correlated with the estimated transmitral flow) In some cases, a consideration of the transmitral gradient, the total cardiac output, and the state of the other valves of the heart as determined clinically and by combined catheterization and ventriculography, may be necessary even to determine which lesion is responsible for the presenting symptoms

Because nearly complete (albeit temporary) relief of symptoms will follow a relatively small enlargement of the functional valve area, in the past many surgeons have not felt it necessary to persist in mobilizing the valve to the extent necessary for the complete elimination of the pressure gradient and the prevention of refusion

A recent review[2] of 598 of our patients operated on for mitral stenosis five or more years ago has shown that 32 (5 per cent) already have been submitted for reoperation for recurrence of the mitral obstruction (Table 1) We are convinced that these 32 patients represent merely a fraction of those who either have already or will in time develop restenosis Such is the ultimate outcome to be expected in individuals who have been treated by the older operative technics in which only an in-

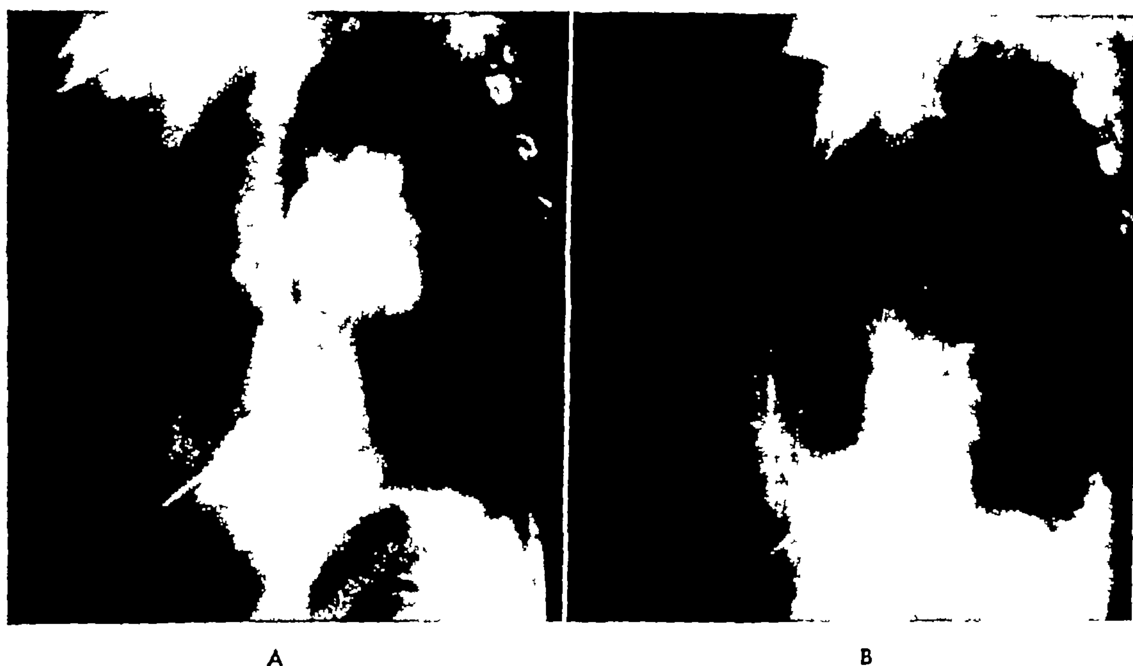


Fig 14 Pre- and postoperative left ventriculograms showing A incompetent mitral valve, and B same valve restored to near competence (Nichols, H T J Thoracic Surg, 33 102 (Jan) 1957)

complete mobilization of the valve has been accomplished.

Restenosis nearly always is the result of inadequate surgical mobilization of the valve. It has not, in our experience, been associated with proven recurrent rheumatic activity[3,4,5] although, of course, this must be considered an ever-present possibility

Severe Calcification: Severe calcification of the valve is not a contraindication to surgery although such calcification may limit the degree of functional restoration which can be obtained. Similarly advanced age no longer is considered a contraindication to operation.

Pregnancy: Mitral valve surgery may be carried out during pregnancy with every expectation of an excellent result, especially if it is performed during the middle trimester In the first trimester there is some risk of miscarriage and during the last trimester the burden on the heart and circulation is at its peak.

Discussion

Surgical relief of mitral stenosis became a reality in 1948[1] with the establishment of the concept that the stenosis must be relieved without the creation of regurgitation. In terms of surgical technic, this concept usually has been interpreted to mean that the surgeon must limit himself to separating the valve components only within the *lines of commissural fusion.* The theoretic basis for such a "mitral commissurotomy" procedure was decribed in detail (although under another term) with regard to pertinent anatomic and pathologic aspects in the excellent paper of Brock[6] in 1952 Mitral valve surgery either without or with instrumental help now has been practiced for more than a decade. However the "standard" technic has been found wanting in many respects.

(1) The operative mortality for mitral commissurotomy has remained formidable, ranging between 5 and 8 per cent for most large series. This, of course, is influenced to a considerable extent by the functional status of the patient, but even in the best type of patient, the risk of mortality has not been negligible

(2) The results of mitral commissu

TABLE 1

Percentage of Patients with Recurrent Stenosis Among All Survivors for Each Calendar Year of Our Operative Intervention for Predominant* Mitral Stenosis

Year of Initial Operation	*Number of Survivors*	*Number of Patients with Later Recurrence*	*Percentage of Patients with Recurrence*
1918	2	1	50
1919	12	1	8
1930	78	5	6
1931	208	10	5
1932	998	15	5
Total for first five years (1918-1932)	598	[illegible]	5

(Bailey C. P and Morse D P J Internat Coll Surg 31 8 Jan 1959)

rotomy while definite in terms of subjective improvement[4, 7, 8] have been clearly disappointing from the objective point of view. In the great majority of cases the objective signs that led to the preoperative diagnosis of mitral stenosis have remained even after the operation.

Physiologic studies, while showing definite improvement[9, 10] after operation, are not consistently satisfactory.

(3) The risk of serious morbidity following the operation, particularly that due to operatively produced mitral regurgitation and that due to arterial embolization resulting from dislodgement of thrombotic material from the left atrium during the transappendageal manipulations, has remained uncomfortably high.

(4) Recurrence of symptomatology after a period of improvement lasting one or more years following surgery is becoming increasingly evident. Statistics on the incidence of restenosis after mitral commissurotomy differ widely[2, 11] but there cannot be any doubt today that it is a very common occurrence and will, in our opinion, become even more evident with the passage of the years. It is obvious, therefore, that the procedure of mitral commissurotomy in its original sense cannot be looked upon as a satisfactory operation for the relief of mitral stenosis, especially since a much superior operation is available today. The considerable merit of mitral commissurotomy lies in the fact that it was the first major breakthrough in the development of intracardiac surgery. As the result of the enormous experience accumulated in this field during the last few years, however, a more profound anatomic and physiologic understanding has evolved and this has led to fundamental changes in the concepts and technics concerned with the surgical correction of mitral stenosis.

A very major portion of the operative mortality associated with mitral commissurotomy, as well as the not inconsiderable number of cases in which the surgeon must be satisfied with compromise results, is related to the fact

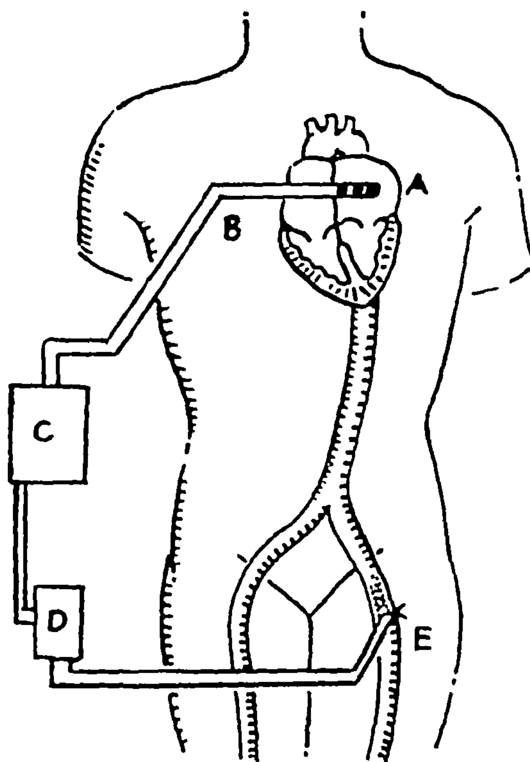


Fig 15 Diagrammatic representation of selective left ventricular bypass for support of the circulation during a closed operation for mitral stenosis. The oxygenated blood from the left atrium is drained off by a catheter introduced trans septally and then is returned to the arterial circuit by way of one of the femoral arteries. Thus only the mitral valve and the left ventricle are bypassed.

that the function of the left ventricle which is essential for moment-to-moment survival of the organism is interfered with by the surgical manipulations at the valve. Although, in the majority of cases, the heart is able to stand this degree of insult, one can clearly perceive that it is quite impossible to predict the cardiac reaction in any individual instance. It would appear, therefore, that some form of

temporary support of the circulation is most desirable during the manipulations on the valve.

Support by Extracorporeal Bypass Circulatory support by subtotal or partial bypass is utilized in certain clinics during closed surgery for mitral stenosis. In open procedures for this condition physiologic support is part and parcel of the method. In the better risk patient only a selective left ventricular bypass is advised, the oxygenated blood from the left atrium being pumped into a systemic artery (Fig 15) In individuals with evidence or history of persistent right ventricular failure or extreme pulmonary hypertension, and in most elderly patients, it is preferable to bypass the whole heart

This requires the interposition of an oxygenator into the extracorporeal by pass circuit (Fig 16) The surgical manipulations are generally still performed with a closed technic since the mortality in this very ill type of patient is high with open heart surgery The total bypass set up does, of course, provide conditions for completely open mitral valve surgery Such an open technic is being used more freely as the safety of open procedures increases. When one expects to find excessive pathologic change in the valve, the presence of a loose clot in the atrium, or an element of concomitant mitral insufficiency true "open heart" surgery should always be practiced Then the left thoracic approach and wide inci-

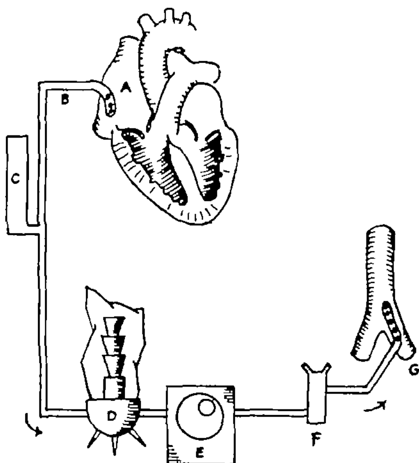


Fig 16. Bypass of the entire heart is preferred in extremely sick individuals, and especially in those with advanced pulmonary vascular changes. This method lends itself to circulatory support with either an open or a closed operative technic.

sion of the left atrium facilitate the exposure and technical opening of the valve

It is now realized generally that mere division of the commissural scar tissue cannot be expected to restore the mitral valve to normal or even near normal function Since even a comparatively small enlargement of the valve orifice frequently will produce a considerable improvement in the patient's condition (at least temporarily), the limitations of the original surgical concept of mitral commissurotomy have not always been obvious in clinical practice From an extensive personal and group experience in the quest for better immediate results and more persistent long-term improvement, a surgical technic of *mitral valve reconstitution* based upon a completely different concept has been evolved Since in the great majority of cases the valve mechanism will have been destroyed irretrievably and since the carnified mural portion usually cannot be rendered mobile to any significant degree, a method has been worked out whereby, by "neostrophingic" mobilization,[12, 13] one may fashion a new "septal leaflet" out of the anterior aspect of the scarred valve "cone" The resultant mechanism is very different from that of the normal valve "cylinder" but the created valve flap becomes displaced in effortless fashion into the left ventricular cavity during diastole and impinges effectively upon the shelf-like mural ridge during the subsequent systole The newly created leaflet hinges upon its characteristically uninvolved thin and pliable basal tissue This new valve mechanism is established by extending the separation of the valve tissue through and well beyond the commissural scars in lines projected toward the trigonal terminations of the horseshoe shaped mitral valve "ring" (Fig. 17). Since under conditions of artificial (mechanical) support of the circulation the surgeon is able to proceed deliberately and without haste, he can methodically and, little by little, cleave the valve in such a way as to mobilize this tongue-like flap without the creation of any significant degree of regurgitation.

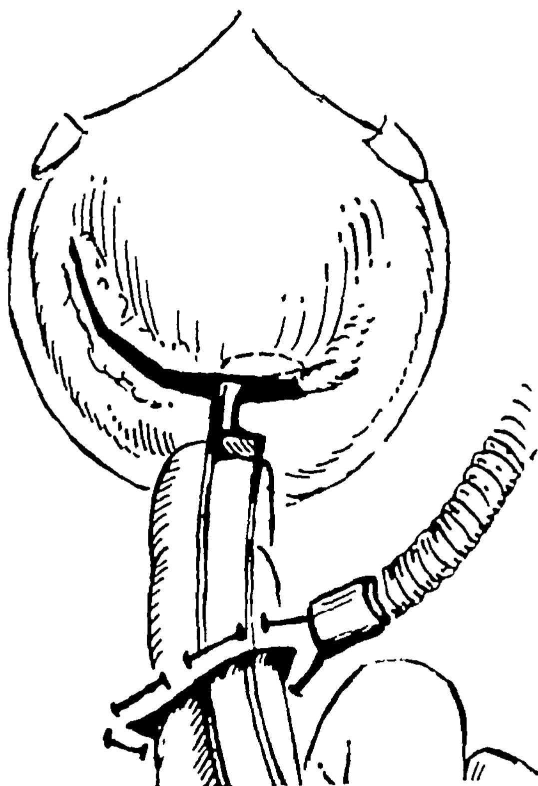


Fig 17 Neostrophingic mobilization of the stenotic mitral valve being carried out by a closed technic

Anatomic Approach

The anatomic approach provided by the left atrial appendage or nearby atrial wall has a number of serious drawbacks in closed (digitally controlled) surgery of the mitral valve The manipulations of the valve from this avenue of approach are rather difficult especially with regard to the right or "posteromedial" commissure because of the direction of the left atrioventricular channel (Fig 18) which can be reached only "around the corner" The friability of the left atrial appendage

provides an added hazard to entrance by this route. Futhermore, in the presence of left atrial thrombosis the site of which most commonly is the lumen of the appendage and perhaps also the nearby atrial wall, manipulation through this thrombus-bearing area obviously may very easily lead to operative arterial embolization.

The anatomic approach through the

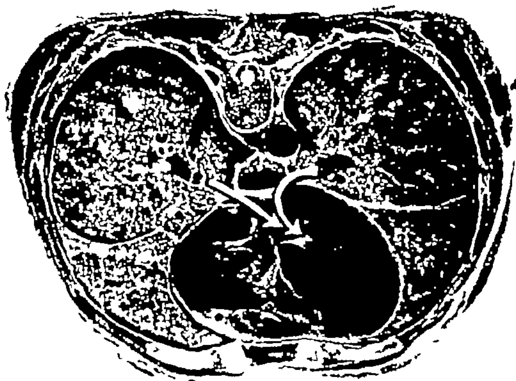


Fig. 18: Cross section of thorax at level of the left atrioventricular channel. Note contrasting directions of approach which explain the technical advantages of the right thoracic route in closed procedures for mitral stenosis. This does not hold in open procedures. (Seara P.: Estudio Anatomico Topografico del Thorax y del Abdomen (1944) Buenos Aires.)

(right) interatrial groove, therefore, is far superior to the left appendageal one for a closed operative procedure. When an open approach is chosen, however the left lateral approach and a wide incision of the left atrium provides extremely satisfactory guidance by ocular means for proper valve mobilization. The nonfunctioning heart readily may be displaced or distorted to bring the valve into clear view. Furthermore, venting of the left ventricle is easily accomplished with the left lateral approach while it is extremely difficult from the right (Fig. 19). This "venting" by permitting continuous and complete evacuation of any entrapped air from the left ventricle throughout the procedure effectively precludes the development of air embolization. A further advantage accrues when the left lateral approach is chosen for open-heart operations. This is related to the accessibility to the left ventricle and the facility with which defibrillation or rhythmic compression may be applied if necessary.

The Evolution of Mitral Surgery

Developmental Stages: Surgery for mitral stenosis has gone through at least three major phases since the first suc-

cessful mitral commissurotomy operation was performed in 1948[1] In the initial phase (approximately the first three to four years) the valve was incised, "fractured," or dilated as a crude surgical feat without full realization of the importance of opening both commissures fully or of separating adequately any crossfused subvalvular structures (chordae tendineae) In the second phase of surgical development from 1953 to 1957, more rational concepts especially as to technic of mobilization were worked out A bicommissural operation was attempted in all cases and the "neostrophingic" procedure[12, 13, 14] was devised whereby a tongue-like flap was created from the longer "septal" side of the valve sleeve While this amounted to the substitution of an entirely new type of valve mechanism for the natural one, it was not technically feasible in all clinical cases It proved to be an effective mechanism, and the resulting valve orifice and functional capacity approach practically normal values This concept of complete functional mobilization was extended to include extensive separation of the subvalvular structures However, in some patients it was not possible to so mobilize the stenotic valve even with the technically more advantageous right-sided approach, because of spells of severe hypotension or the frank inability of the heart to withstand prolonged intracardiac manipulations Only recently has it become possible to open the valve completely in practically every patient At the present time the heart may be supported by (1) a selective left ventricular bypass (valve opened by closed technic) with a right-sided approach, or (2) total heart-lung bypass (valve opened by closed technic) or, (3) by using complete heart-lung bypass and a fully open technic with a left-sided approach Since September 1959, the authors have employed this last technic in practically all of their patients. Any of these technics may be chosen, and with artificial circulatory support will permit unhurried work upon the valve so that the ultimate goal of the surgery may be attained in practically every case

Modern Surgical Technics

Right-Sided Approach: ***Closed Operation:*** The operation by way of a dissection of the interatrial sulcus (patient in the supine position) not only allows the surgeon to work with equal facility at both extremities of the valve, but also avoids the inherent risk of particulate embolization which attends digital entrance through the most frequently involved sites for thrombosis (the left atrial appendage and adjacent lateral atrial wall) With the growth and evolution of more advanced technical concepts and the consequent elevation of standards of mobilization, the frequency of instrumentation of the valve has increased steadily over the years until now some instrumental aid is used in more than 80 per cent of the patients

Open Operation: With the increasing safety of extracorporeal bypass procedures and the progressive improvement in the special equipment utilized, it is logical to presume that a completely open technic will be used more and more frequently in the surgical correction of mitral stenosis

Indeed, it seems only reasonable to anticipate that, as the safety of the open procedure becomes equal to and finally surpasses that of the closed method in basic safety (with regard to simple survival), the closed method will be supplanted completely It would seem that the obvious technical advantages of

the open method and the consistently superior type of valve mobilization obtainable by it (especially in the hands of those who are less experienced in the *closed technics*) make the open-heart procedure the method of choice.

We have come to the conclusion that the open technic should certainly be used whenever it is anticipated that unusual technical difficulties may be encountered during the definitive part of the procedure (history of previous *operation for mitral stenosis*, *evidence* of extensive calcification of the valve by radiographic means, the presence of associated mitral insufficiency the likelihood of intra atrial thrombosis—suggested by history of repeated embolic phenomena, etc.) In addition, since the younger and more vigorous subject tolerates *open heart* bypass extremely well, we have begun to carry out open procedures for mitral valve lesions in such patients. As yet we have not lost a single patient in this category and in each an extremely good technical mobilization has been achieved without the creation of any significant element of incompetence

On the other hand, in the very ill patient, especially the one with severe, long standing pulmonary hypertension and chronic congestive heart disease, it is felt that present-day open heart technics, in themselves, impose a significant additional element of morbidity and mortality Hence many such patients are still being operated on by us by a closed technic using the right sided approach.

When the open technic is chosen, for a number of reasons it seems preferable to choose the left posterolateral thoracic approach even in those who have been operated on previously through a left transpleural incision The reasons may be stated as follows

1 Only by way of a left thoracic approach is the left ventricle fully visualized so that it can be "vented" adequately and reliably enough to preclude the possibility of arterial air embolism, be compressed manually if required, and be defibrillated electrically if necessary

2 Intermittent or temporary clamp occlusion of the pulmonary artery and/or aorta is facilitated, a most practical point since at least a minor degree of aortic incompetence coexists in approximately one-half of these patients.

3 Excellent visualization of the atrial aspect of the valve is readily obtainable with an open technic utilizing this approach since the non-*functioning heart may be dislocated in any* desired direction

4 The "target points" (trigonal terminations of the horseshoe shaped annulus fibrosus) are readily delineated from this vantage point permitting critical accuracy in establishing the proper cleavage lines.

5 Once the valve substance has been divided both chordopapillary systems come into view so that they may be mobilized or split appropriately in such a way as to provide maximal mobility to the created tongue like valve flap while, at the same time maintaining maximal support of the mobilized structure

6 Any associated element of incompetence is both recognizable (by closed pre bypass exploration of the left atrial lumen with a finger) and readily correctible during the "open" phase of the procedure.

Needless to say, the open technic facilitates the mechanical removal of any intra atrial thrombus and the piece meal (usually) extraction of calcific encrustations from the valve. Any inadvertently dislodged calcific fragments may be picked up individually with fine thumb forceps.

The necessary oxygenator bypass system may be connected by double cannulation of the femoral veins and arteries. Should the venous return prove insufficient, by this technic, to provide the necessary flow to maintain the circulation at normothermic temperatures, a degree of hypothermia may be induced early by the use of a heat-exchanger to reduce the flow requirements of the body to that obtain

able by the extracorporeal system Following completion of the definitive procedure and after restoration of an effective intrinsic circulatory mechanism, the heat-exchanger is utilized to rewarm the patient. This femoral vascular "hook-up" has the advantage that the perfusion circuit is established and subsequently removed with the patient in the more "physiologic" supine position Should severe hypotension or embarrassment of cardiac action supervene when the patient is being turned into the lateral position for operation (or at the conclusion during the converse procedure), the pump oxygenator may be started promptly as a highly effective supportive or even resuscitative measure while the intended operative program is continued in methodical fashion

In the more vigorous subject, in whom such an untoward episode would seem most improbable, the thoracic incision may be completed before heparinization, then after its establishment the right ventricular outflow tract (or main pulmonary artery) and the descending thoracic aorta may be cannulated directly. Normal flow can then be provided at normothermic temperatures

Regardless of the technic of cannulation used, complete cardiopulmonary bypass (at the selected temperature level) should be established The left ventricle should be "vented" surgically before opening the left atrium We have found the insertion of a two inch segment of a plastic multifenestrated #30 F. catheter into the lumen, by way of a stab wound made in the subapical region of the ventricle, a most satisfactory method of "venting" (Fig 19) A suture uniting the catheter segment to the ventricular wall will prevent inadvertent dislodgement Blood which escapes from the tube may be aspirated from the pericardial sac by the "low-pressure" suction device and returned to the extracorporeal circuit At the conclusion of the definitive procedure, and after the left atrial wall has been repaired securely in two layers, the left atrium is compressed to evacuate any entrapped air. Then the catheter segment is removed and the ventriculotomy wound is repaired with sutures, after any residual intraventricular air has been expelled by the ventricular contractions or by manual compression of the heart

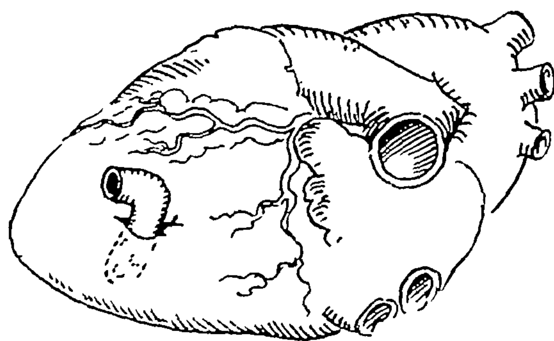


Fig 19 Method of "venting" the left ventricle As long as the ventricular chamber is in free communication with the atmospheric air sufficient pressure cannot be generated to overcome the intracardiac blood pressure Thus, the ever present possibility of air embolism is precluded

The mitral valve is visualized most advantageously by making a long incision in the left atrial wall just posterior to the appendage and the coronary sinus The lips of the opening may be separated by placement of traction sutures or by the use of nontraumatic clamps The atrium is evacuated of any thrombotic material present and the valve orifice is examined The "trigonal" or "target points" are located by inspection and palpation The diminutive valve orifice is lengthened by incising the indurated tissues at its extremities in the direction of the "target points," taking due care to pre-

serve proper chordopapillary support for both the septal flap (which is being created) and for the residual mural "shelf" Once both cleavage lines have reached flexible valve tissue, provided that a free and adequate orifice has been established the tissue separation is terminated lest an element of incompetence be produced

The supporting chordae may be found joined in a short "yoke just beneath the valve orifice. If so the "stem" of the Y which really is the thickened papillary muscle, is split longitudinally by terminal incision at its bifurcation followed by end-on digital pressure fully to its origin from the ventricular wall

Any loosely attached calcific material found upon the valve margins is removed and the flexibility of the mobilized flap is tested by instrumental manipulation. If an element of insufficiency sufficient to cause disability has preexisted it may be repaired at this time. In idiopathic cases the incompetence usually will respond to a technic of "shortening of the annulus fibrosus (preferably by actual division and imbrication) or to application of prosthetic material to the retracted mural margin of the valve. Usually the regurgitation, in these cases, will take place in the region of the posteromedial pole of the valve.

In patients whose valves have been rendered incompetent by the trauma of previous (closed) operations for mitral stenosis there may be found any one of several varieties of damage Chordae tendineae, or one of the papillary muscles may have been divided continuity of the valve margin may have been interrupted at an incorrect point a valvular perforation may have been created The specific damage and the appropriate corrective measure usually will be self evident and repair may be undertaken at once.

The use of the left-sided approach for closed commissurotomy for mitral stenosis without the aid of any form of bypass hardly seems justifiable at this time. It is becoming ever more evident that, whenever possible, operations for mitral stenosis should be performed only at the larger heart centers where a considerable volume of such surgical material and comprehensive facilities are available. It is only under these conditions that sufficient technical skill can be developed by the operating team so that optimal results with this technically difficult valve (both with respect to its critical location and the usually advanced pathologic change) can be assured

Special Technical Considerations

Emboli The interatrial sulcus approach to the valve from the right provides relatively safe entrance for "closed heart" surgery even in patients with extensive left atrial thrombosis. We have never found clotted material actually adherent to or covering this site in our entire operative experience (in contrast, the left atrial appendage contains clot in 25 to 30 per cent of patients with mitral stenosis) Consequently the incidence of fatal and nonfatal operative embolism has been drastically reduced with the exclusive utilization of the right sided approach whenever a closed procedure is chosen (Table 2)

Nevertheless, the presence of a free floating ball valve thrombosis or of a long pedicled finger of fresh clot which may be encountered immediately above the valve necessitates immediate surgical removal One of the "Litwak" aspirating tips[13] connected through large bore tubing to a source of strong

TABLE 2

Incidence of Fatal and Nonfatal Operative Embolization by Different Technics[13]

	Mitral Commissurotomy From the Left Side (811 cases*)		Neostrophingic Mobilization From the Right Side (227 cases)	
	Nonfatal	*Fatal*	*Nonfatal*	*Fatal*
Cerebral	14	16	3	2
Extremity	7	3	2	
Pulmonary	2	6		
Visceral	3	1		
Cerebral plus one of above	1	2		
Total	27	28	5	2
	(3 3%)	(3.5%)	(2 2%)	(0 9%)

* As previously reported Bailey, C P Surgery of the Heart Lea & Febiger, Philadelphia, 1955, p 544

suction will permit efficient removal of the clot (Fig 20)

Upon completion of the definitive surgery, it is customary at our clinic to establish a small (8 to 10 mm), communicating, slit-like, atrial septal defect

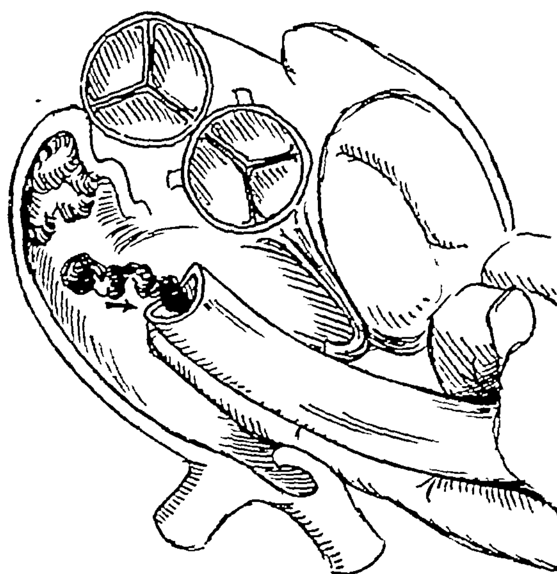


Fig 20 Removal of free floating or loosely attached thrombotic material from the left atrium when operating for mitral stenosis by a closed technic One of the Litwak suction tips of appropriate size is introduced into the left atrium along the palmar surface of the operating finger Its tip is brought into proximity to the clot At a given signal the clot is aspirated along with considerable blood

to serve as a "blow-off" should excessive left atrial pressure develop during the immediate postoperative period When severe mitral stenosis has been present for years, the left ventricle will have become hypotrophic and may be unable to adjust immediately to the larger volume load permitted by relief of its inlet obstruction The created defect, which is made in the dorsal muscular portion of the atrial septum, has been shown on repeat catheterization to have become obliterated within a few weeks

As mentioned, the incidence of operative embolization has been reduced to a new low level since the right-sided approach has been used in closed operations for mitral stenosis (Table 2)

Atrial Fibrillation: This may occur postoperatively in a previously rhythmic patient and is best controlled by digitalization In the majority of instances, it will revert to a regular rhythm within a few weeks

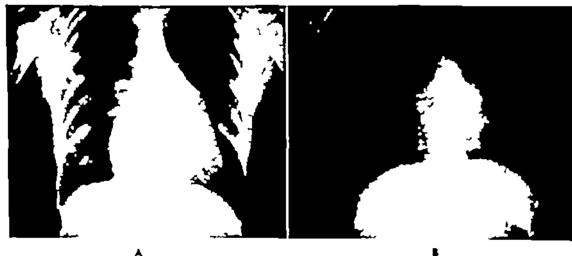
The so-called "post-commissurotomy syndrome" consisting of fever, pain in

the chest, and pleural or pericardial effusion may occur after any intrapericardial operation including those for the repair of congenital defects. It is now clearly apparent that it does not represent a rheumatic reactivation[7, 13] The fever will respond to the aspiration of any accumulated serosanguinous fluid from the pleural or pericardial sacs and to the administration of an anti-inflammatory (anti-stress) drug of the corticoid group

Results

Mortality: The mortality rate in surgery for mitral stenosis depends upon the functional classification and physiologic state of the patients presented for operation. Mortality figures presented from thirty seven outstanding clinics in a recent international survey[7] vary from less than 2 per cent in patients who are minimally symptomatic to more than 25 per cent in those who have chronic intractable failure Among 10 000 operations for mitral stenosis there were approximately 7 per cent operative deaths. The overall mortality involved in treating all types of patients (including many of the worst risks) by our group has been reduced by the measures described to less than 5 per cent. It is anticipated that with further improvements in the technics of application of extracorporeal bypass it will be possible to bring nearly all of the most critically ill and many frankly terminal patients through the operation successfully

The former two great risks of hypotension with circulatory failure, and inadvertent creation of mitral insufficiency have been decreased greatly by the use of supportive bypass which permits much more methodical exploration of the valve and unhurried instrumentation



A B

Fig 21 Reduction in the overall size of the heart following relief of the mitral obstruction. A. Preoperative posteroanterior roentgenogram, B postoperative comparable roentgenogram six months followup.

Follow Up Status of Postoperative Patients The patients operated on with the closed neostrophingic technic (right thoracic approach) in the second phase of the surgery (second four years) for mitral stenosis have shown abolition of the mitral diastolic murmur in 50 per cent of cases as compared with 8 to 10 per cent in patients operated from the left side during the first stage (1948 1953) It is expected that the increasingly frequent recognition of recurrent stenosis (already as high as 12

TABLE 3

Pre- and Postoperative Catheterization Data in Mitral Stenosis[9]

		Cardiac Output (L/min)	*Valve flow (cc/V.F.P sec *)*	*Mitral Gradient (mm Hg **)*	*Mitral Valve Area (cm²)*
Case II					
1	Before	4 4	145	15	1.2
	After	6 4	296	5	4 3
2	Before	4 1	110	11	1 0
	After	3.3	188	3	3 8
3	Before	3 9	125	20	0 8
	After	3 8	226	4	3 6

*V F P = Ventricular filling phase
**Pressure gradient measured by planimetry

per cent in our earlier cases) will be much reduced with our present more adequate procedures Those patients who do not have excessively enlarged hearts or irreversible pulmonary vascular change and, in particular, those in whom the ventricles are still in good working order may be restored clinically to a near-normal condition, with every expectation that they will remain in this state for a great many years

The physiologic evidence of the efficiency of the newer operation is dramatic The mitral gradient is reduced greatly or actually abolished, the cardiac output, the valve flow and, consequently, the valve area as determined by catheterization are all increased to near-normal figures (Table 3). X-ray studies (Fig 21) confirm the long-term improvement To date none of the patients operated on by the neostrophingic method has returned with refusion (three year followup) The older type of "closed heart" operation continues to eventuate in an incomplete relief of the obstructive lesion and, consequently, in recurrence of the obstruction in a high percentage of patients

References

1. Bailey, C P Dis Chest, 15 377, 1949
2. Bailey, C P and Morse, D P J Internat Coll Surg, 31 8, 1959
3. Lisan, P., Reale, A, and Likoff, W Ann Int Med., 50 1352, 1959
4. Likoff, W and Uricchio, J F J A M.A., 166 737, 1958
5. Brest, A N, Uricchio, J, and Likoff, W Ibid., 171 249, 1959
6. Brock, R C Brit Heart J, 14 489, 1952
7. Surgical Treatment of Mitral Stenosis Rep Sect. Cardiovas Surg Am Coll Chest Physicians Dis of Chest, 35 435, 1959
8. Conklin, W S, Maurice, G L, and Bergquist, S F Dis of Chest, 35 291, 1959
9. Dickens, J, Villaca, L, Woldow, A., and Goldberg, H Brit Heart J, 19 419, 1957
10. Dogliotti, A M, Actis-Dato, A, and Gentilli, R Surgery, 44 706, 1958
11. Patterson, G C and Marshall, J R Brit Heart J, 21 174, 1959
12. Bailey, C P and Hirose, T J Thoracic Surg, 35 5, 1958
13. Bailey, C P., Hirose, T, and Morse, D P Am J Cardiol., 1 91, 1958
14. Bailey, C P and Morse, D P J Thoracic Surg, 33 427, 1957

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